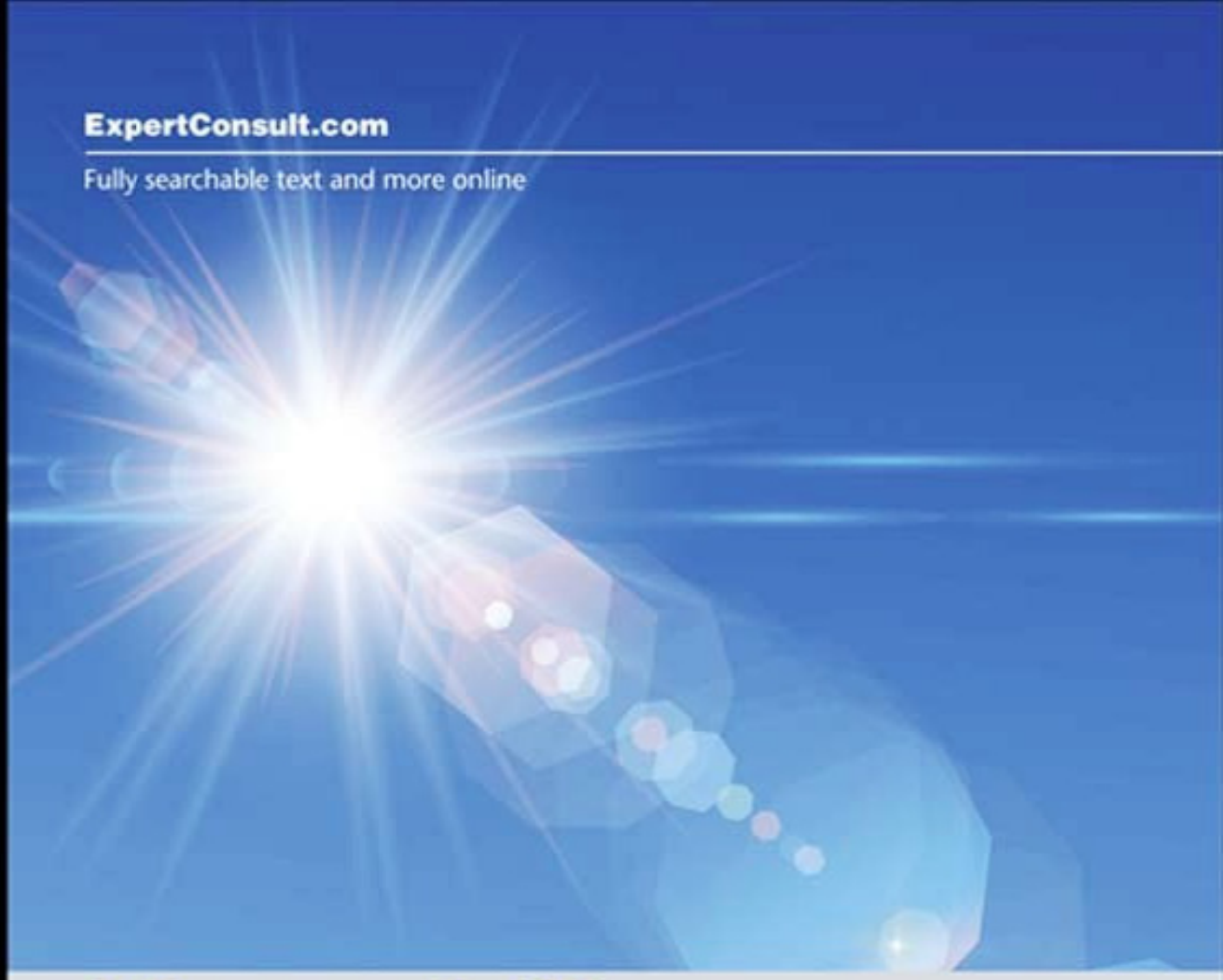

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A SYSTEMATIC APPROACH

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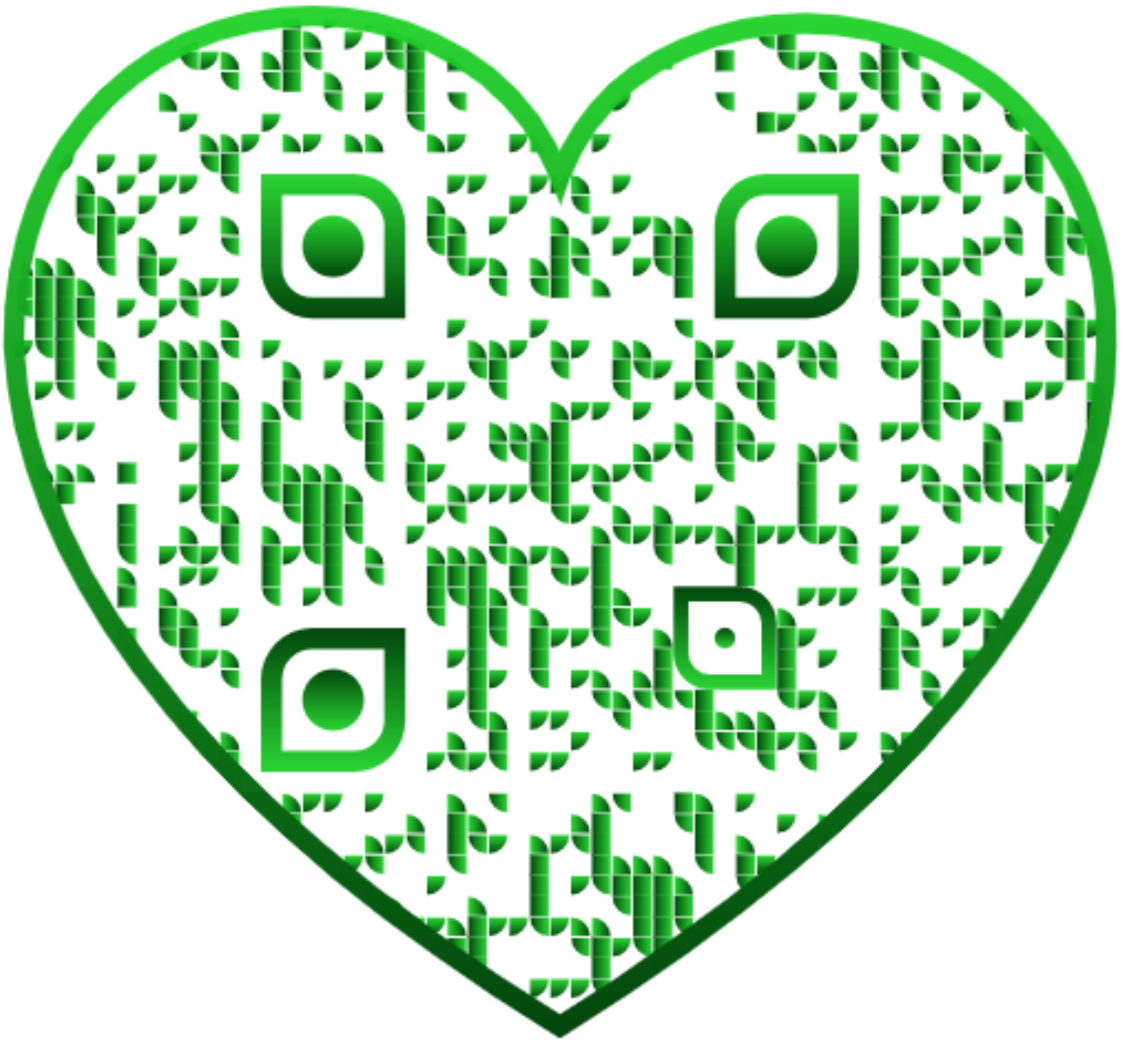

Kanski's Clinical Ophthalmology

A SYSTEMATIC APPROACH

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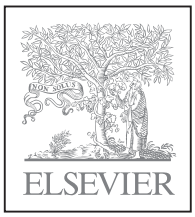

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INTRODUCTION

Anatomy

The skin (Fig. 1.1A) consists of the epidermis, dermis and related structures (adnexa).

Epidermis

The epidermis is comprised of four layers of keratin-producing cells (keratinocytes). It also contains melanocytes, Langerhans cells and Merkel cells. The layers of the epidermis around the eye are described below; cells migrate superficially, undergoing maturation and differentiation through successive layers.

- **Keratin layer** (stratum corneum or horny layer) consists of flat cells devoid of nuclei.
- **Granular cell layer** (stratum granulosum) typically consists of one or two layers of flattened cells containing keratohyaline granules.
- **Prickle cell layer** (stratum spinosum) is approximately five cells deep. The cells are polygonal in cross-section and have abundant eosinophilic cytoplasm. Their free borders are united by spiny-appearing desmosomes (cellular junctions).
- **Basal cell layer** (stratum basale) comprises a single row of columnar-shaped proliferating cells containing melanin derived from adjacent melanocytes.

Dermis

The dermis is much thicker than the epidermis. It is composed of connective tissue and contains blood vessels, lymphatics and nerve

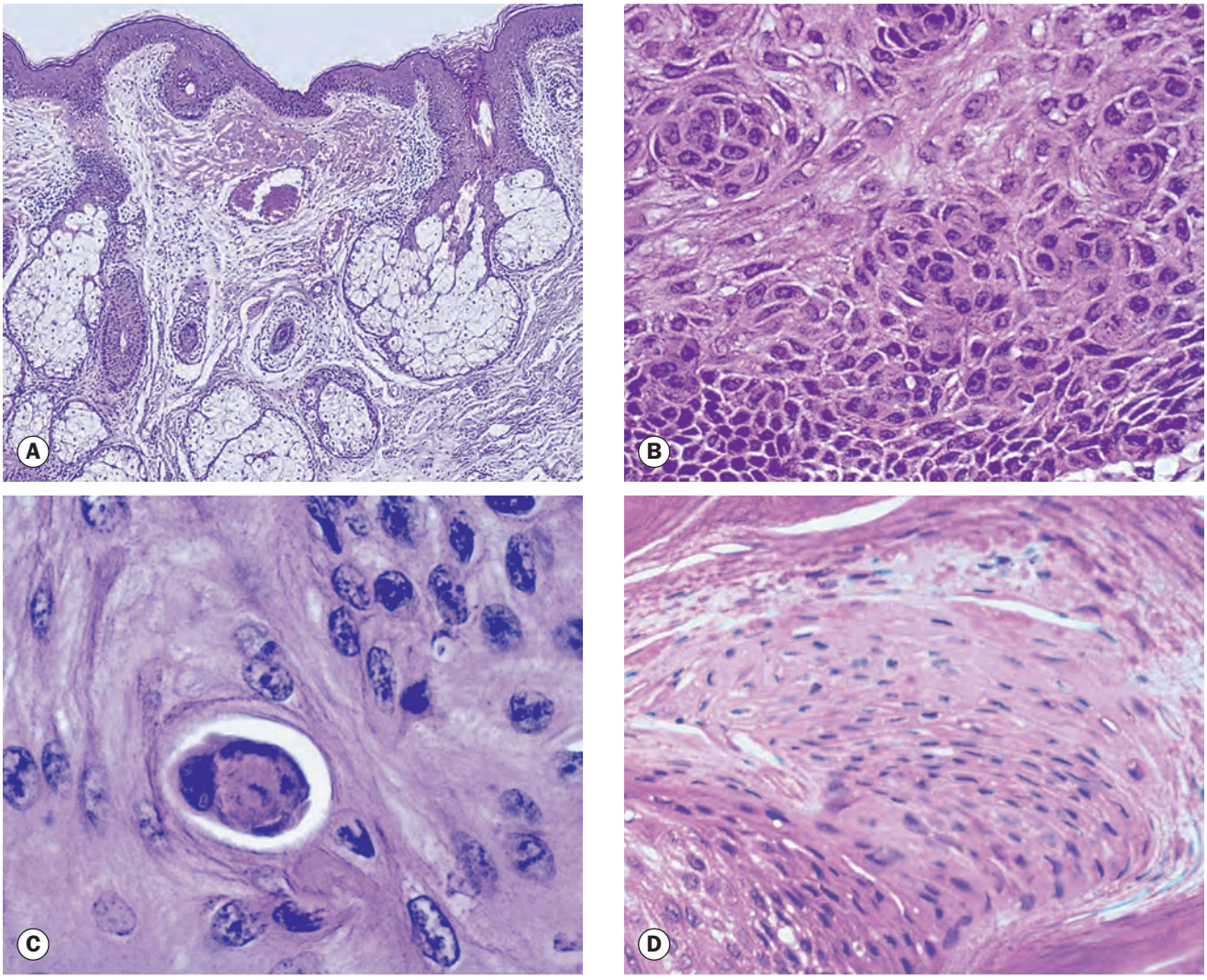


Fig. 1.1 Eyelid skin. **(A)** Normal skin is composed of keratinized stratified epithelium that covers the surface; pilosebaceous elements are conspicuous in the dermis and a few blood vessels and sweat glands are also seen; **(B)** dysplasia with loss of cell polarity; **(C)** dyskeratosis – a non-surface epithelial cell producing keratin; **(D)** parakeratosis – retention of cell nuclei into the surface keratin layer

(Courtesy of J Harry – fig. A; J Harry and G Misson, from Clinical Ophthalmic Pathology, *Butterworth-Heinemann, 2001 – figs B–D)*

fibres in addition to fibroblasts, macrophages and mast cells; upward dermal extensions (papillae) interdigitate with downward epidermal projections (rete ridges). In the eyelid the dermis lies on the orbicularis muscle. Adnexa lie deep in the dermis or within the tarsal plates.

- **Sebaceous glands** are located in the caruncle and within eyebrow hairs. Tiny sebaceous glands are associated with the thin (vellus) hairs covering periocular skin.
- **Meibomian glands** are modified sebaceous glands found in the tarsal plates. They empty through a single row of 20–30 orifices on each lid. A gland consists of a central duct with multiple acini, the cells of which synthesize lipids (meibum) that form the outer layer of the tear film.
- **Glands of Zeis** are modified sebaceous glands associated with lash follicles.
- **Glands of Moll** are modified apocrine sweat glands opening either into a lash follicle or directly onto the anterior lid margin between lashes; they are more numerous in the lower lid.
- **Eccrine sweat glands** are distributed throughout eyelid skin and are not confined to the lid margin, in contrast to glands of Moll.
- **Pilosebaceous units** comprise hair follicles and their sebaceous glands (see Fig. 1.1A).

Terminology

Clinical

- **Macule.** Localized area of colour change without infiltration, depression or elevation, less than 1 cm in diameter.
- **Papule.** A solid elevation less than 1 cm in diameter.
- **Vesicle.** Circumscribed lesion containing serous fluid; less than 0.5 cm across.
- **Bulla.** A large (more than 0.5 cm) serous fluid-filled lesion; plural – bullae.
- **Pustule.** A pus-filled elevation less than 1 cm in diameter.
- **Crust.** Solidified serous or purulent exudate.
- **Nodule.** A palpable solid area measuring more than 1 cm.
- **Cyst.** A nodule consisting of an epithelial-lined cavity filled with fluid or semi-solid material.
- **Plaque.** A solid elevation of the skin, greater than 1 cm in diameter.
- **Scale.** Readily detached fragments of shed keratin layer.
- **Papilloma.** A benign neoplastic warty or tag-like projection of the skin or mucous membrane.
- **Ulcer.** A circumscribed area of epithelial loss; in skin an ulcer extends through the epidermis into the dermis.

Histological

- **Tumour** strictly refers only to a swelling, though is commonly used to denote a neoplasm.
- **Neoplasia.** Abnormal tissue growth, either benign (localized, non-invasive and non-spreading) or malignant (progressive growth with the potential for distant spread).
- **Atypia** refers to an abnormal appearance of individual cells, e.g. abnormal mitotic figures.
- **Dysplasia** is an alteration of the size, morphology and organization of cellular components of a tissue. There is disturbance of normally structured and recognized layers of tissue (e.g. loss of cell polarity – Fig. 1.1B).
- **Carcinoma *in situ*** (intraepidermal carcinoma, Bowen disease) exhibits dysplastic changes throughout the thickness of the epidermis.
- **Hyperkeratosis.** An increase in thickness of the keratin layer that appears clinically as scaling. Hyperkeratosis can be a feature of benign or malignant epithelial tumours.
- **Acanthosis.** Thickening of the prickle cell layer.
- **Dyskeratosis** is keratinization other than on the epithelial surface (Fig. 1.1C).
- **Parakeratosis** is the retention of nuclei into the keratin layer (Fig. 1.1D).

General considerations

- **Classification.** Epidermal, adnexal or dermal.
- **Diagnosis.** The clinical characteristics of benign lesions are a tendency to a lack of induration and ulceration, uniform colour, limited growth, regular outline and preservation of normal lid margin structures. Biopsy may be required if the appearance is suspicious.
  - Incisional biopsy involves removal of a portion of a lesion for histopathology.
  - Excision biopsy is performed on small tumours and fulfils both diagnostic and treatment objectives.
- **Treatment** options include:
  - Excision of the entire lesion and a small surrounding portion of normal tissue.
  - Marsupialization involves the removal of the top of a cyst allowing drainage of its contents and subsequent epithelialization.
  - Ablation with laser or cryotherapy.

NON-NEOPLASTIC LESIONS

Chalazion

Pathogenesis

A chalazion (meibomian cyst) is a sterile chronic granulomatous inflammatory lesion (lipogranuloma) of the meibomian, or sometimes Zeis, glands caused by retained sebaceous secretions. Histopathology shows a lipogranulomatous chronic inflammatory picture with extracellular fat deposits surrounded by lipid-laden epithelioid cells, multinucleated giant cells and lymphocytes (Fig. 1.2A). Blepharitis is commonly present; rosacea can be associated with multiple and recurrent chalazia. A recurrent chalazion should be biopsied to exclude malignancy.

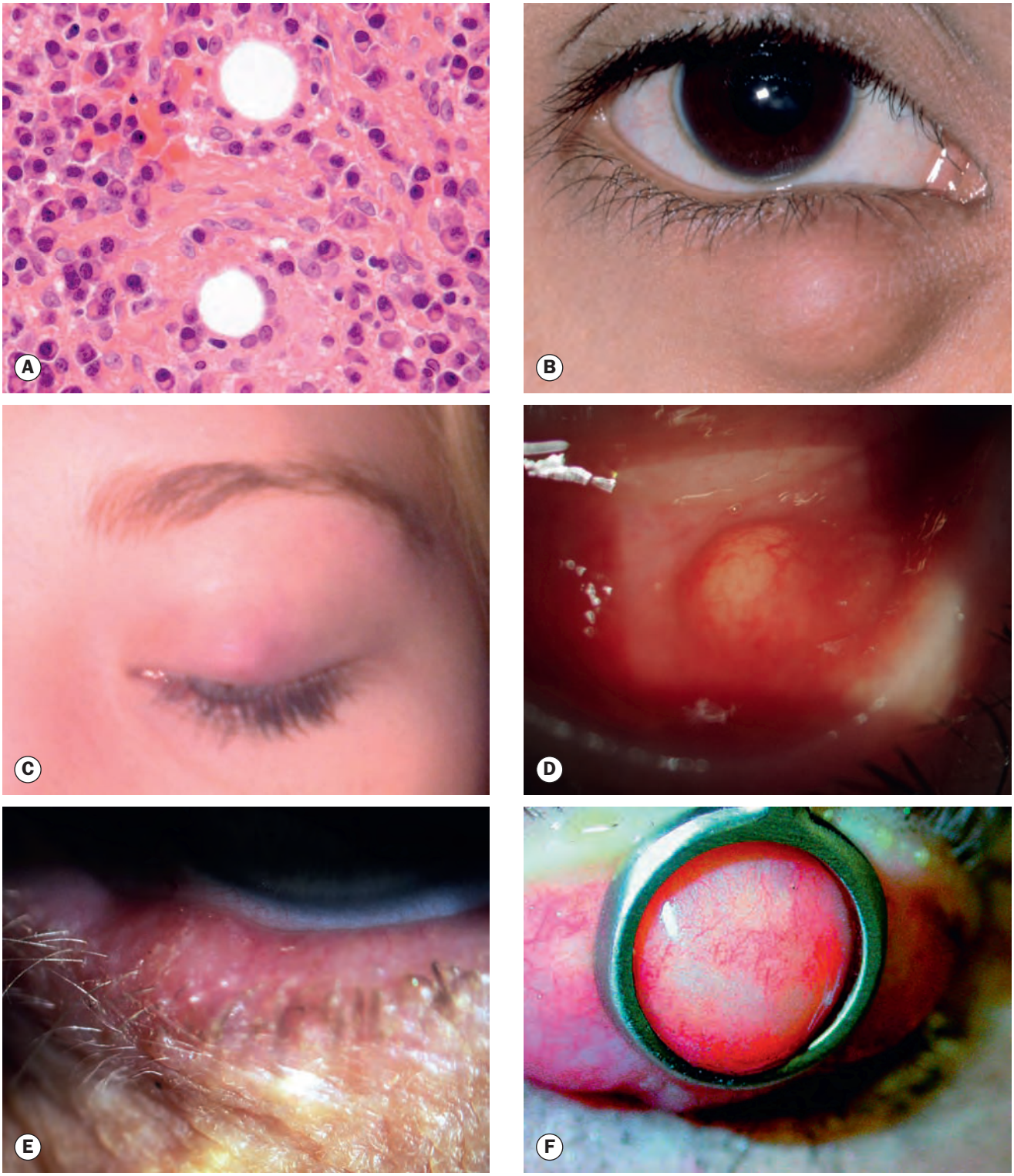


Fig. 1.2 Chalazion. **(A)** Histopathology shows a lipogranuloma; the large pale cells are epithelioid cells and the well-demarcated empty space contained fat dissolved out during processing; **(B)** uninflamed chalazion; **(C)** acutely inflamed lesion; **(D)** conjunctival granuloma; **(E)** marginal chalazion; **(F)** conjunctival view of chalazion clamp in place prior to incision and curettage

(Courtesy of J Harry and G Misson, from Clinical Ophthalmic Pathology, *Butterworth-Heinemann 2001 – fig. A; J Nerad, K Carter and M Alford, from 'Oculoplastic and Reconstructive Surgery', in* Rapid Diagnosis in Ophthalmology, *Mosby 2008 – fig. F)*

Diagnosis

- **Symptoms**
  - Subacute/chronic: gradually enlarging painless rounded nodule (Fig. 1.2B).
  - Acute: sterile inflammation or bacterial infection with localized cellulitis (Fig. 1.2C); differentiation may be difficult. A secondarily infected meibomian gland is referred to as an internal hordeolum.
- **Signs**
  - A nodule within the tarsal plate, sometimes with associated inflammation.
  - Bulging inspissated secretions may be visible at the orifice of the involved gland.
  - There may be an associated conjunctival granuloma (Fig. 1.2D).
  - A lesion at the anterior lid margin – a marginal chalazion (Fig. 1.2E) – may be connected to a typical chalazion deeper in the lid or be due to isolated involvement of a gland of Zeis.

Treatment

- **Oral antibiotics** are required for significant bacterial infection, but not for sterile inflammation.
- **Conservative.** At least a third of chalazia resolve spontaneously so observation may be appropriate, especially if the lesion is showing signs of improvement, though early definitive treatment has been reported to lead to higher patient satisfaction.
- **Hot compress** application several times daily may aid resolution, particularly in early lesions.
- **Expression.** Compression between two cotton-tipped applicators is sometimes effective in expressing the contents of a fresh lesion near the lid margin.
- **Steroid injection** into or around the lesion has been reported to give similar resolution rates to incision and curettage (see below). It may be preferred for marginal lesions or lesions close to structures such as the lacrimal punctum because of the risk of surgical damage.
  - Reported regimens include 0.2–2 ml of triamcinolone acetonide aqueous suspension diluted with lidocaine to a concentration of 5 mg/ml, and 0.1–0.2 ml of 40 mg/ml, injected with a 27- or 30-gauge needle.
  - The success rate following one injection is about 80%; a second can be given 1–2 weeks later.
  - Local skin depigmentation and fat atrophy are potential but uncommon complications, the risk of which may be reduced by avoidance of infiltration immediately subcutaneously or by utilizing a conjunctival approach.
  - Retinal vascular occlusion has been described as a complication, probably due to intravascular injection with subsequent embolization.
- **Surgery**
  - Following local anaesthesia infiltration, the eyelid is everted with a specialized clamp (Fig. 1.2F), the cyst is incised vertically through the tarsal plate and its contents curetted.
  - Limited excision of solid inflammatory material (sent for histopathology) with fine scissors may be helpful in some cases, especially if there is no focus of secretions.
  - A suture should not be used.
  - Topical antibiotic ointment is used three times daily for 5–7 days following curettage.
- **Marginal lesions** can be managed by steroid injection, by curettage of an associated deeper chalazion, by shave curettage or by incision and curettage via a horizontal incision on the conjunctival surface or vertically through the grey line.
- **Prophylaxis**
  - Treatment of blepharitis, e.g. daily lid hygiene regimen.
  - Systemic tetracycline may be required as prophylaxis in patients with recurrent chalazia, particularly if associated with acne rosacea.

Other eyelid cysts

- **Cyst of Zeis** is a small, non-translucent cyst on the anterior lid margin arising from obstructed sebaceous glands associated with the eyelash follicle (Fig. 1.3A).
- **Cyst of Moll** (apocrine hidrocystoma) is a small retention cyst of the lid margin apocrine glands. It appears as a round, non-tender, translucent fluid-filled lesion on the anterior lid margin (Fig. 1.3B).
- **Sebaceous (pilar)** cyst is caused by a blocked pilosebaceous follicle and contains sebaceous secretions; the gland orifice will often be visible (Fig. 1.3C). It is only rarely found on the eyelid although it may occasionally occur at the inner canthus.
- **Comedones** are plugs of keratin and sebum within the dilated orifice of hair follicles that often occur in patients with acne vulgaris. They may be either open (blackheads) containing a darkened plug of oxidized material (Fig. 1.3D), or closed (whiteheads).
- **Milia** are caused by occlusion of pilosebaceous units resulting in retention of keratin. They are tiny, white, round, superficial papules that tend to occur in crops (Fig. 1.3E).
- **Epidermal inclusion** cyst is usually caused by implantation of epidermis into the dermis following trauma or surgery. It is a slow-growing, round, firm, superficial or subcutaneous lesion containing keratin (Fig. 1.3F).
- **Epidermoid** cyst is uncommon and usually developmental, occurring along embryonic lines of closure. It is similar in appearance to an epidermal inclusion cyst.
- **Dermoid** cyst is usually subcutaneous or deeper and is typically attached to the periosteum at the lateral end of the brow (Fig. 1.3G). It is caused by skin sequestered during embryonic development.
- **Eccrine hidrocystoma** is less common but similar in appearance to a cyst of Moll except that it is usually located along the medial or lateral aspects of the lid, and is close to but does not involve the lid margin itself (Fig. 1.3H).

Fig. 1.3 Eyelid cysts. **(A)** Cyst of Zeis; **(B)** cyst of Moll; **(C)** sebaceous cyst; **(D)** comedones – blackheads; **(E)** milia; **(F)** epidermal inclusion cyst;

Xanthelasma

Introduction

Xanthelasma (plural – xanthelasmata) is a common, frequently bilateral condition typically affecting middle-aged and elderly individuals. It is a subtype of xanthoma. Hyperlipidaemia is found in about one-third of patients, in whom corneal arcus may also be present. In contrast to chalazion, fat in xanthelasmata is mainly intracellular, with lipid-laden histiocytes (foam cells) in the dermis (Fig. 1.4A).

Diagnosis

Xanthelasmata are yellowish subcutaneous plaques, usually in the medial aspects of the eyelids (Fig. 1.4B), commonly bilateral and are multiple (Fig. 1.4C).

Fig. 1.3, Continued **(G)** dermoid cyst; **(H)** eccrine hidrocystomas
(Courtesy of A Pearson – figs D, F and H)

Treatment

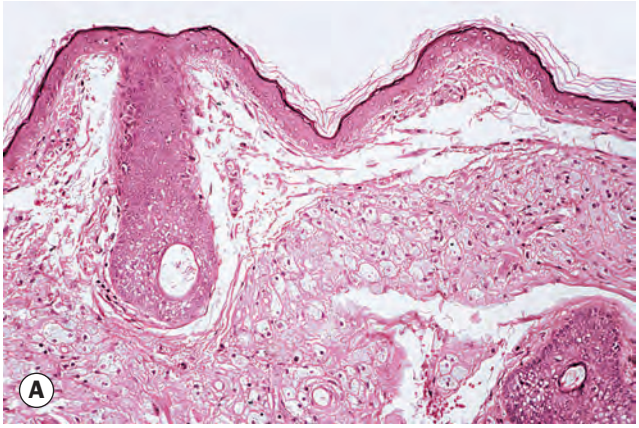
This is principally for cosmesis. Recurrence occurs in up to 50%, and is most common in patients with hypercholesterolaemia.

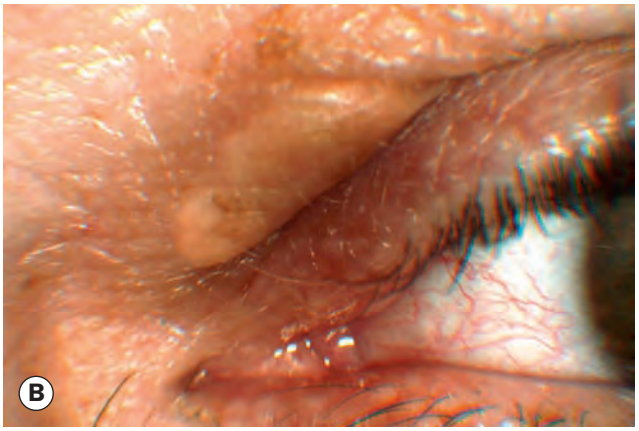
- **Simple excision** is commonly performed where adequate excess skin is present.
- **Microdissection.** Larger lesions can be raised in a flap, the fatty deposits dissected from overlying skin under a surgical microscope using microscissors, and the skin replaced.
- **Other methods.** Good results can be obtained using chemical peeling with bi- or trichloroacetic acid. Laser ablation and cryotherapy have advantages but may be more prone to scarring, including pigmentary changes.

BENIGN EPIDERMAL TUMOURS

Squamous cell papilloma

Squamous cell papilloma is a very common benign epithelial tumour with a variable clinical appearance, including narrow-based (pedunculated or 'skin tag' – Fig. 1.5A), pink broad-based





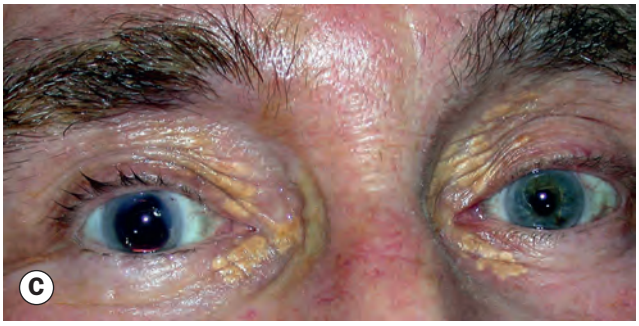


Fig. 1.4 Xanthelasma. **(A)** Histopathology showing foamy histiocytes within the dermis; **(B)** large isolated lesion; **(C)** multiple bilateral smaller lesions
(Courtesy of J Harry – fig. A; S Chen – fig. C)

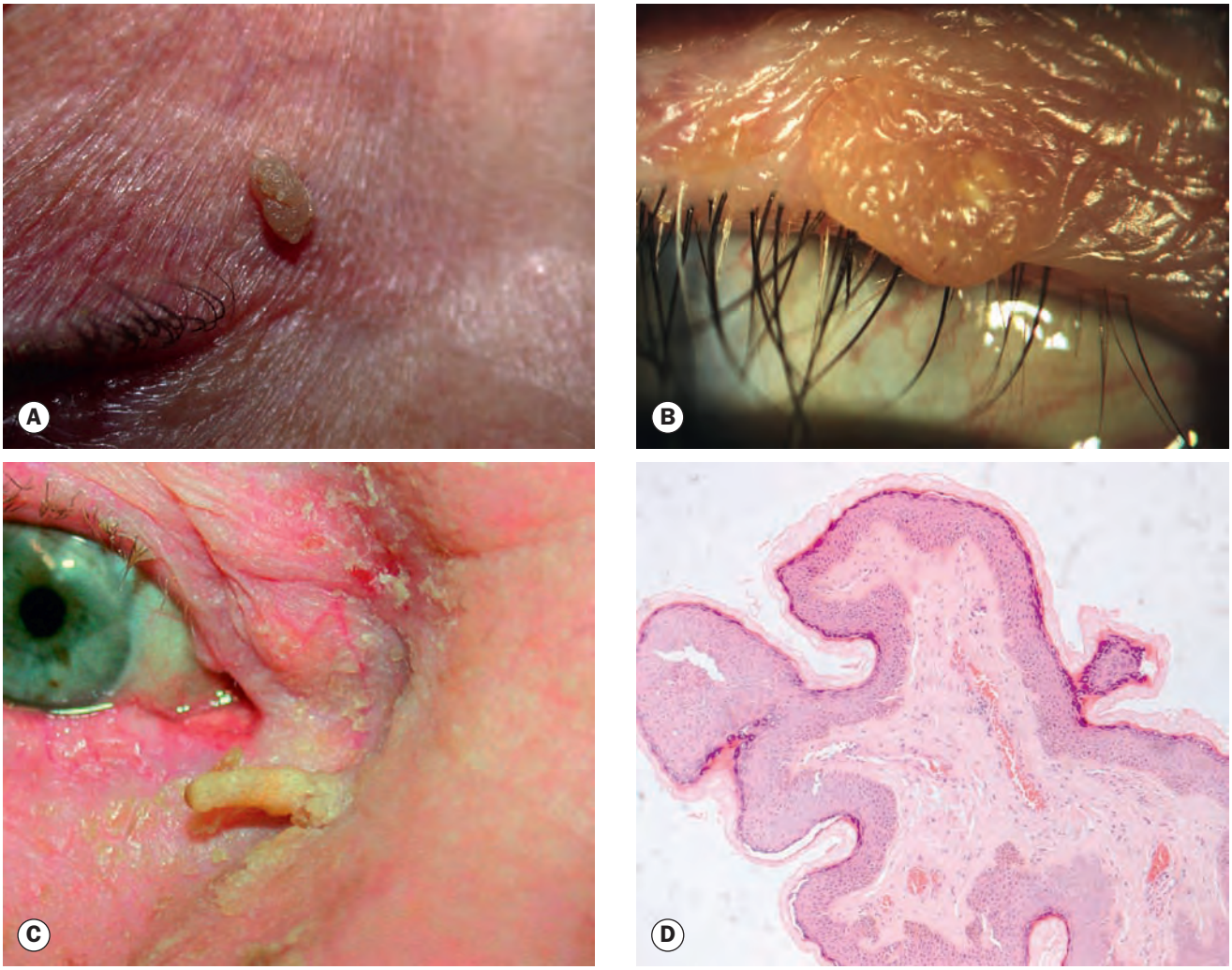


Fig. 1.5 Squamous cell papilloma. **(A)** Pedunculated 'skin tag'; **(B)** sessile lesion; **(C)** hyperkeratotic filiform lesion; **(D)** histopathology shows finger-like projections of fibrovascular connective tissue covered by irregular acanthotic and hyperkeratotic squamous epithelium
(Courtesy of A Pearson – fig. C; J Harry – fig. D)

(sessile – Fig. 1.5B) and whitish thread-like (filiform) hyperkeratotic lesions similar to a cutaneous horn (Fig. 1.5C). Histopathology in all clinical types is similar, showing finger-like projections of fibrovascular connective tissue covered by irregular acanthotic and hyperkeratotic squamous epithelium (Fig. 1.5D). The incidence increases with age; at least some cases result from human papilloma virus infection. Treatment usually involves simple excision, but other options include cryotherapy and laser or chemical ablation.

Seborrhoeic keratosis

Seborrhoeic keratosis (basal cell papilloma) is an extremely common slowly growing lesion found on the face, trunk and extremities of elderly individuals as a discrete light- to dark-brown plaque with a friable, greasy, verrucous surface and a 'stuck-on' appearance (Fig. 1.6A). They are frequently numerous. The differential diagnosis includes pigmented basal cell carcinoma, naevus and melanoma. Histopathology shows expansion of the squamous epithelium of the epidermis by proliferating basal cells, sometimes with keratin-filled horns or cystic inclusions (Fig. 1.6B). Treatment involves shave biopsy (occasionally simple excision), electrodesiccation with curettage, laser ablation, cryotherapy with liquid nitrogen, and chemical peeling.

Actinic keratosis

Actinic (solar, senile) keratosis is a common slowly growing lesion that rarely develops on the eyelids. It typically affects elderly, fair-skinned individuals on areas of sun-damaged skin such as the forehead and backs of the hands, and appears as a hyperkeratotic plaque with distinct borders and a scaly surface that may become fissured (Fig. 1.7A). Occasionally the lesion is nodular or wart-like and may give rise to a cutaneous horn. Histopathology shows irregular dysplastic epidermis with hyperkeratosis, parakeratosis and cutaneous horn formation (Fig. 1.7B). It has potential, though

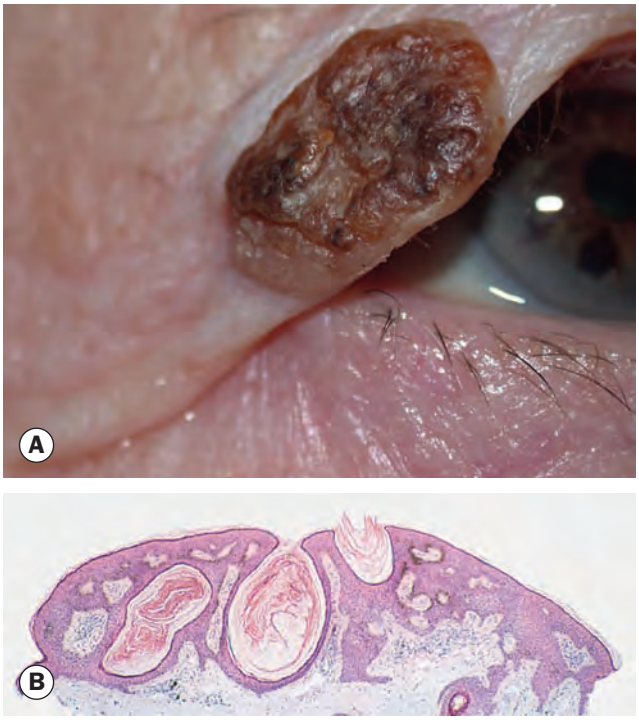


Fig. 1.6 Basal cell papilloma. **(A)** Typical 'stuck-on' appearance; **(B)** histopathology showing an elevated expansion of the epidermis with proliferation from basal cells – horn cysts and pseudohorn cysts are present
(Courtesy of A Pearson – fig. A; J Harry – fig. B)

low, for transformation into squamous cell carcinoma. Treatment involves biopsy followed by excision or cryotherapy.

BENIGN PIGMENTED LESIONS

Freckle

A freckle (ephelis, plural ephelides) is a small (generally 1–5 mm) brown macule due to increased melanin in the epidermal basal layer, typically in sun-exposed skin (Fig. 1.8); numbers vary with the level of sun exposure and can sometimes regress completely. Histopathology shows hyperpigmentation of the basal layer of the epidermis, with a normal melanocyte population.

Congenital melanocytic naevus

Congenital naevi are uncommon and histologically resemble their acquired counterparts (see below). They are usually small and of uniform colour. Rare variants include a 'kissing' or split naevus that involves the upper and lower eyelid (Fig. 1.9A) and may occasionally contain numerous hairs (Fig. 1.9B), and a very large lesion covering an extensive area of the body ('giant hairy naevus' – Fig. 1.9C). Large lesions have the potential for malignant transformation (up to 15%). Treatment, if necessary, involves complete surgical excision.

Acquired melanocytic naevus

Diagnosis

The clinical appearance and potential for malignant transformation of naevi are determined by their histological location within the skin.

- **Junctional** naevus occurs in young individuals as a uniformly brown macule or plaque (Fig. 1.10A). The naevus cells are located at the junction of the epidermis and dermis and have a low potential for malignant transformation (Fig. 1.10B).
- **Compound** naevus occurs in middle age as a raised papular lesion. The shade of pigment varies from light tan to dark

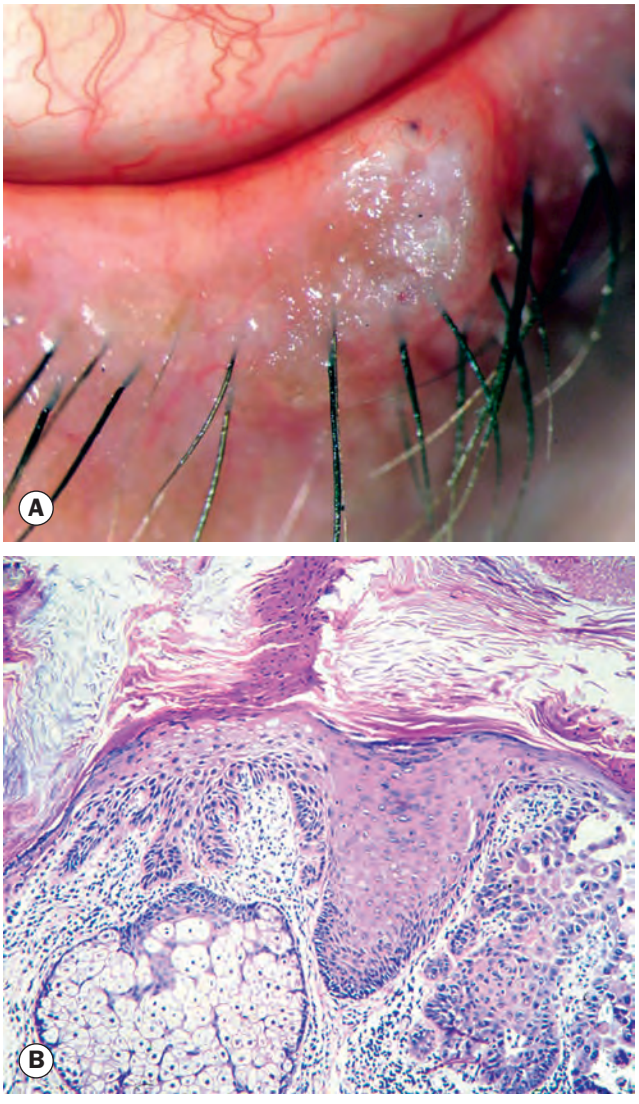


Fig. 1.7 Actinic keratosis. **(A)** Clinical appearance; **(B)** histopathology shows irregular dysplastic epidermis with hyperkeratosis, parakeratosis and cutaneous horn formation
(Courtesy of M Jager – fig. A; J Harry and G Misson, from Clinical Ophthalmic Pathology*, Butterworth-Heinemann 2001 – fig. B)*

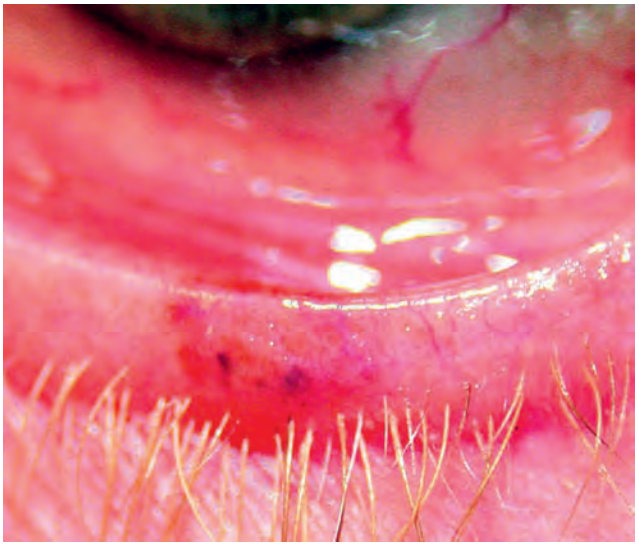


Fig. 1.8 Freckle (ephelis)

brown but tends to be relatively uniform throughout (Fig. 1.10C). The naevus cells extend from the epidermis into the dermis (Fig. 1.10D). It has a low malignant potential related to the junctional component.

- **Intradermal** naevus, the most common, typically occurs in older patients. It is a papillomatous lesion, with little or no pigmentation (Fig. 1.10E). Histologically, naevus cells are confined to the dermis and have essentially no malignant potential (Fig. 1.10F).
- **Variants** of naevi include balloon cell naevi, halo naevi, Spitz naevi (juvenile melanomas) and dysplastic naevi (atypical moles). Multiple dysplastic naevi constitute the dysplastic naevus syndrome (atypical mole syndrome – AMS). Individuals with AMS are at increased risk of developing conjunctival and uveal naevi and cutaneous, conjunctival and uveal melanomas.

Treatment

Treatment is indicated for cosmesis or for concern about malignancy. Excision should be complete in most cases, with at least a 3 mm margin if melanoma is strongly suspected.

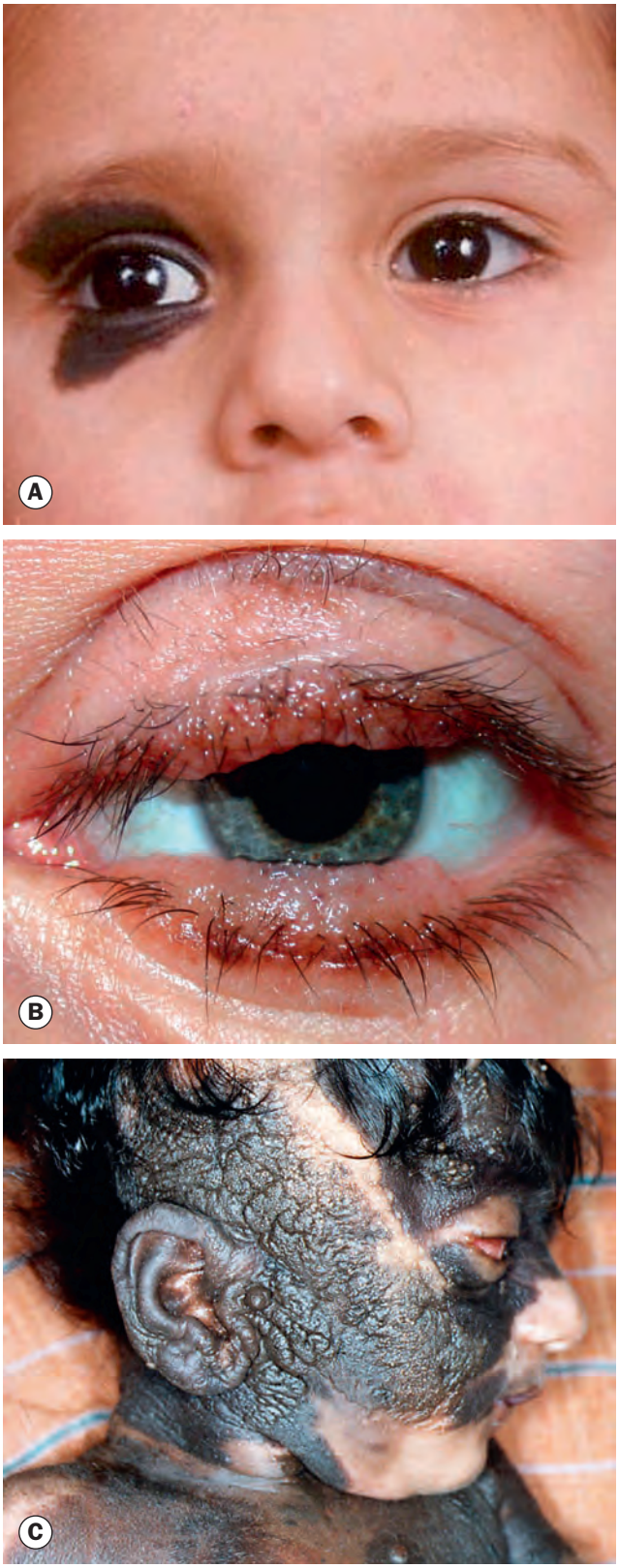


Fig. 1.9 Congenital melanocytic naevus. **(A)** Split naevus; **(B)** split naevus containing hair; **(C)** extensive cutaneous involvement

(Courtesy of A Pearson – fig. B; U Raina – fig. C)

BENIGN ADNEXAL TUMOURS

Syringoma

Syringomas are benign proliferations arising from eccrine sweat glands. They are characterized by small papules that are often multiple and bilateral (Fig. 1.11).

Pilomatricoma

Pilomatricoma (pilomatrixoma, calcifying epithelioma of Malherbe) is derived from the germinal matrix cells of the hair bulb and is the commonest hair follicle proliferation seen by

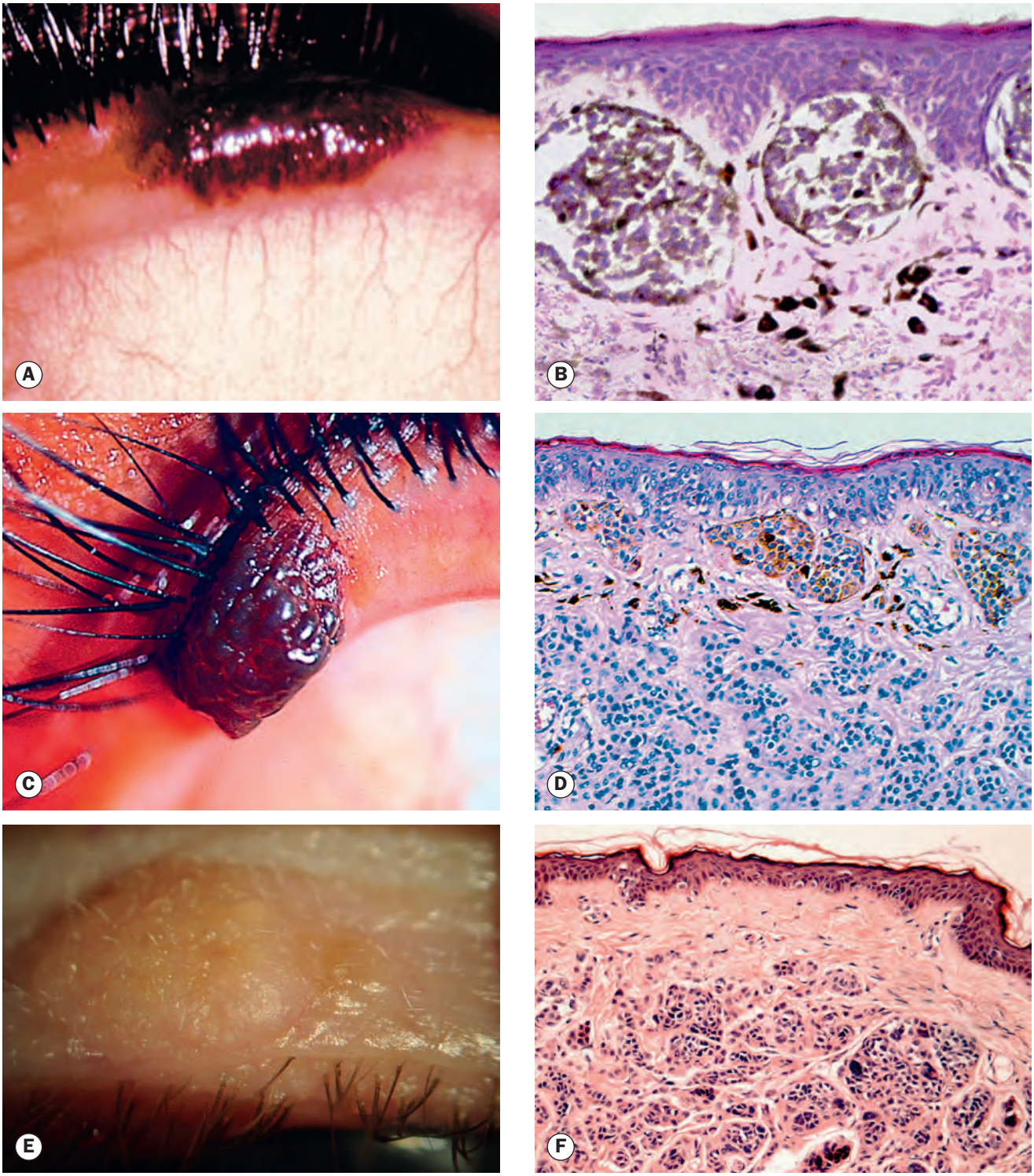


Fig. 1.10 Acquired melanocytic naevus. **(A)** Junctional naevus; **(B)** histopathology shows heavily pigmented naevus cells at the epidermal/dermal junction; **(C)** compound naevus; **(D)** histopathology shows naevus cells both at the epidermal/dermal junction and within the dermis; **(E)** intradermal naevus; **(F)** histopathology shows naevus cells within the dermis separated from the epidermis by a clear zone

(Courtesy of J Harry – figs B, D and F)

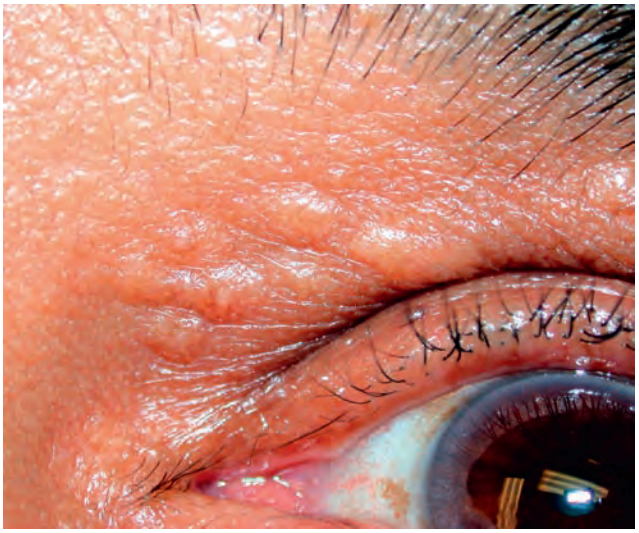


Fig. 1.11 Syringomas
(Courtesy of A Pearson)

ophthalmologists. It affects children and young adults and is more common in females. Clinically it appears as a mobile purplish dermal nodule that may have a hard consistency due to calcification (Fig. 1.12A). Histopathology shows irregular epithelial islands exhibiting viable basophilic cells at the periphery and degenerate 'shadow' cells more centrally (Fig. 1.12B). Calcification is frequently present and there is often a foreign body giant cell reaction. Treatment involves excision. Malignant change is rare. Other, less common, hair follicle proliferations include trichofolliculoma, trichoepithelioma and trichilemmoma.

MISCELLANEOUS BENIGN TUMOURS

Capillary haemangioma

Capillary haemangioma (strawberry naevus) is one of the most common tumours of infancy; it is three times as common in boys as girls. It presents shortly after birth as a unilateral, raised bright red lesion (Fig. 1.13A), usually in the upper lid; a deeper lesion appears purplish (Fig. 1.13B and see also Fig. 3.31). Ptosis is frequent. The lesion blanches on pressure and may swell on crying. There may be orbital extension (see Ch. 3). Occasionally the lesion may involve the skin of the face and some patients have strawberry naevi on other parts of the body. Histopathology shows proliferation of varying-sized vascular channels in the dermis and subcutaneous tissue (Fig. 1.13C). It is important to be aware of an association between multiple cutaneous lesions and visceral haemangiomas, and to consider systemic assessment in appropriate cases. Treatment is described in Ch. 3.

Port-wine stain

Introduction

Port-wine stain (naevus flammeus) is a congenital malformation of vessels within the superficial dermis, consisting histopathologically of vascular spaces of varying calibre separated by thin fibrous septa (Fig. 1.14A). About 10% have associated ocular or CNS involvement, including Sturge–Weber (see below) and other defined syndromes.

Diagnosis

Port-wine stain manifests clinically as a sharply demarcated soft pink patch that does not blanch with pressure, most frequently located on the face. It is usually unilateral and tends to be aligned with the skin area supplied by one or more divisions of the trigeminal nerve (Figs 1.14B and C). Darkening to red or purple takes place with age, and there is commonly associated soft tissue hypertrophy (Figs 1.14D–F). Bleeding may occur from focal overlying lobulations (pyogenic granulomas – see below).

Treatment

Treatment with laser (e.g. pulsed-dye) is effective in decreasing skin discoloration; cosmetically superior results are usually

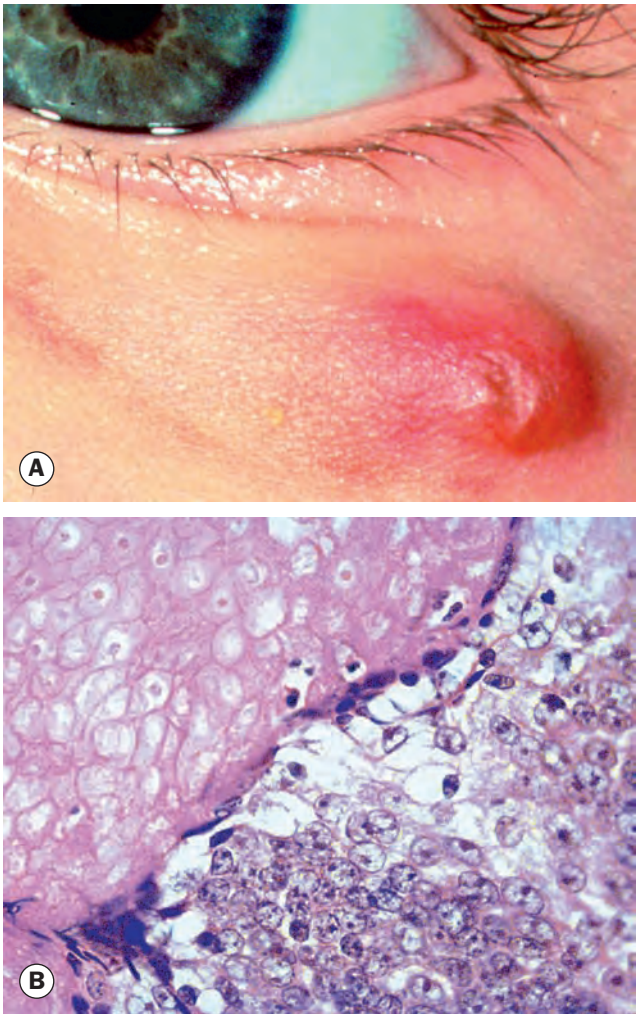


Fig. 1.12 Pilomatricoma. **(A)** Clinical appearance; **(B)** histopathology shows viable basophilic cells to the right and degenerate 'shadow' cells to the left
(Courtesy of J Krachmer, M Mannis and E Holland, from Cornea, *Elsevier 2005 – fig. A; J Harry and G Misson, from* Clinical Ophthalmic Pathology, *Butterworth-Heinemann 2001 – fig. B)*

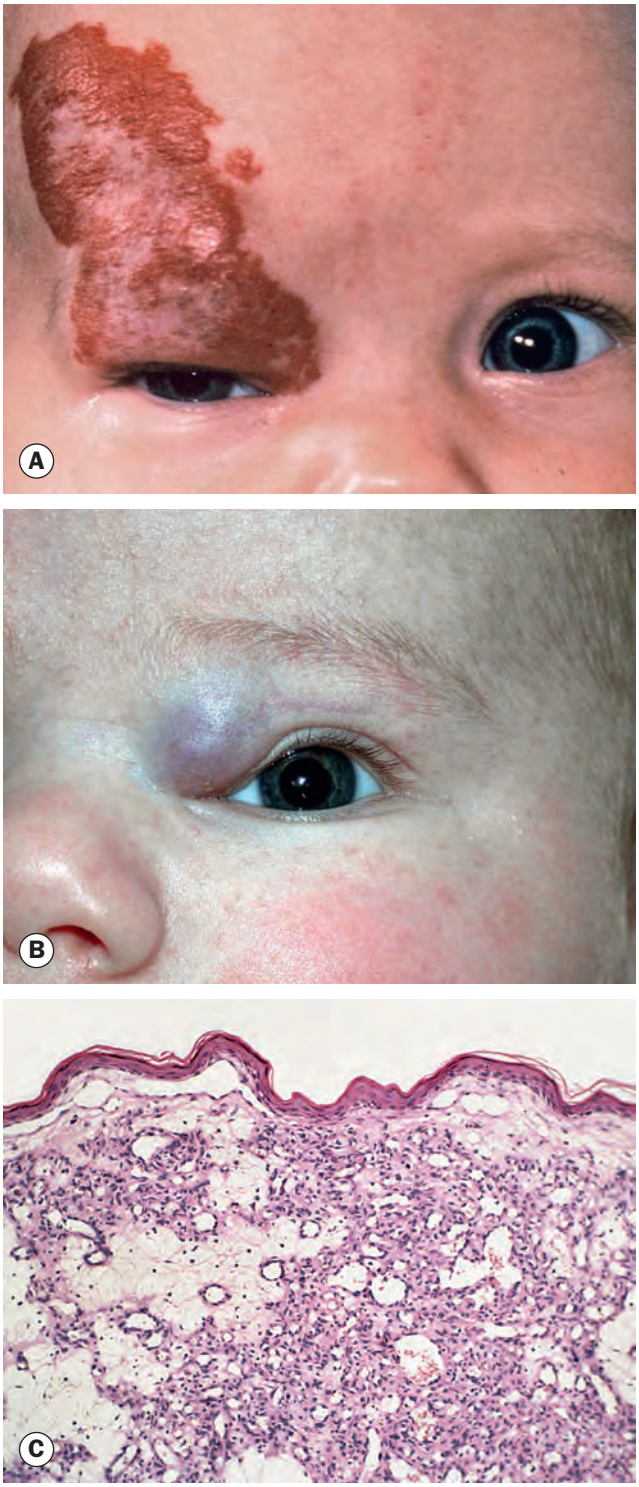


Fig. 1.13 Capillary haemangioma. **(A)** Medium-sized haemangioma; **(B)** mechanical ptosis due to a large lesion; **(C)** histopathology shows vascular channels of varying size within the dermis and subcutaneous tissue
(Courtesy of S Chen – fig. A; J Harry – fig. C)

achieved by early treatment. Topical preparations such as imiquimod and rapamycin, alone or with adjuvant laser, show promise. Soft tissue debulking is used in a small number of cases. Screening for glaucoma should begin in infancy. Systemic investigation is considered in some patients, particularly those with a lesion of the lumbar area.

Sturge–Weber syndrome

Sturge–Weber syndrome (encephalotrigeminal angiomatosis) is a congenital, sporadic phacomatosis.

- **Port-wine stain**, extending over the area corresponding to the distribution of one or more branches of the trigeminal nerve.
- **Leptomeningeal haemangioma** involving the ipsilateral parietal or occipital region may cause contralateral focal or generalized seizures, hemiparesis or hemianopia.
- **Ocular** features may include ipsilateral glaucoma, episcleral haemangioma, iris heterochromia and diffuse choroidal haemangioma (see Ch. 12).

Pyogenic granuloma

Pyogenic granuloma is a rapidly growing vascularized proliferation of granulation tissue that is usually antedated by surgery, trauma or infection, although some cases are idiopathic. Clinically there is a painful, rapidly growing, vascular granulating polypoidal lesion (Fig. 1.15) that may bleed following relatively trivial trauma. Treatment of cutaneous lesions involves excision; conjunctival pyogenic granuloma is discussed in Ch. 5.

Neurofibroma

Cutaneous neurofibromas are benign nerve tumours, usually nodular or pedunculated, that can be found anywhere on the skin. Isolated neurofibromas are common in normal individuals, but if multiple lesions are present neurofibromatosis (see Ch. 19) should be excluded. Plexiform neurofibromas typically present in childhood as a manifestation of neurofibromatosis type 1 with a characteristic S-shaped deformity of the upper eyelid (Fig. 1.16). Treatment of solitary lesions involves simple excision but removal of the more diffuse plexiform lesions may be difficult.

MALIGNANT TUMOURS

The treatment of malignant eyelid tumours in general is discussed at the end of this section.

Rare predisposing conditions

Young patients who suffer from one of the following conditions may develop eyelid malignancies.

- **Xeroderma pigmentosum** is characterized by skin damage on exposure to sunlight, leading to progressive cutaneous abnormalities (Fig. 1.17A). It is inherited in an autosomal recessive (AR) fashion. Affected patients have a bird-like facies and a great propensity to the development of basal cell carcinoma (BCC), squamous cell carcinoma (SCC)

Fig. 1.14 Port-wine stain. **(A)** Histopathology shows widely dilated blood-filled spaces separated by fibrous septa; **(B)** and **(C)** clinical appearance; (**D–F**) progression of port-wine stain over time, with associated underlying soft tissue hypertrophy
(Courtesy of L Horton – fig. A)

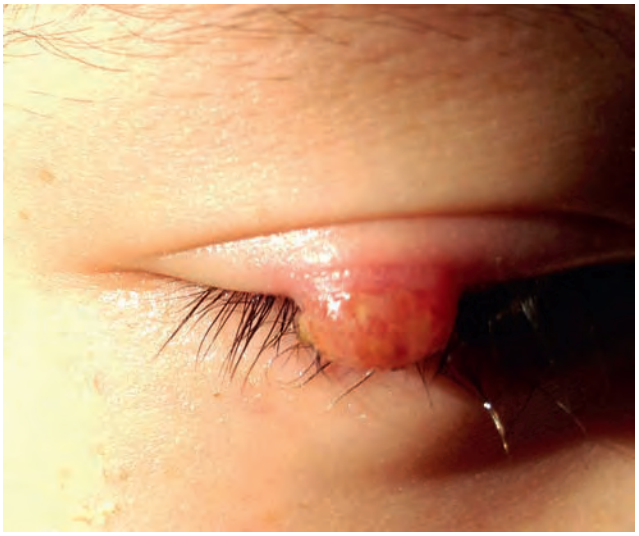


Fig. 1.15 Pyogenic granuloma

Fig. 1.16 Plexiform neurofibroma – characteristic S-shaped upper lid
(Courtesy of J Harry)

Fig. 1.17 Predispositions to eyelid malignancies. **(A)** Xeroderma pigmentosum; **(B)** Gorlin–Goltz syndrome
(Courtesy of J Krachmer, M Mannis and E Holland, from Cornea, *Mosby 2005 – fig. B)*

and melanoma, which are commonly multiple. Conjunctival malignancies have also been reported.

- **Gorlin–Goltz syndrome** (naevoid basal cell carcinoma syndrome) is a rare autosomal dominant (AD) disorder characterized by extensive congenital deformities of the eye, face, bone and central nervous system. Many patients develop multiple small BCC during the second decade of life (Fig. 1.17B) and are also predisposed to medulloblastoma, breast carcinoma and Hodgkin lymphoma.
- **Muir–Torre syndrome** is a rare AD condition that predisposes to cutaneous and internal malignancies. Cutaneous tumours include BCC, sebaceous gland carcinoma and keratoacanthoma. Colorectal and genitourinary carcinomas are the most common systemic tumours.
- **Bazex syndrome** can be used to describe two distinct conditions: (i) Bazex–Dupré–Christol syndrome, an X-linked dominant condition characterized by multiple BCCs, commonly facial including the eyelids, associated with skin changes including follicular indentations without hairs on extensor surfaces (follicular atrophoderma), hypohidrosis and hypotrichosis; (ii) acrokeratosis paraneoplastica of Bazex, in which eczema-like and psoriatiform lesions are associated with an underlying malignancy of the upper respiratory or digestive tract.
- **Other predispositions** include immunosuppression, prior retinoblastoma and albinism.

Basal cell carcinoma

Introduction

BCC is the most common human malignancy and typically affects older age groups. The most important risk factors are fair skin, inability to tan and chronic exposure to sunlight. Ninety per cent of cases occur in the head and neck and about 10% of these involve the eyelid. BCC is by far the most common malignant eyelid tumour, accounting for 90% of all cases. It most frequently arises from the lower eyelid, followed in relative frequency by the medial canthus, upper eyelid and lateral canthus. The tumour is slowly growing and locally invasive but non-metastasizing. Tumours located near the medial canthus are more prone to invade the orbit and sinuses, are more difficult to manage than those arising elsewhere and carry the greatest risk of recurrence. Tumours that recur following incomplete treatment tend to be more aggressive.

Histopathology

The tumour arises from the cells that form the basal layer of the epidermis. The cells proliferate downwards (Fig. 1.18A) and characteristically exhibit palisading at the periphery of a tumour lobule of cells (Fig. 1.18B). Squamous differentiation with the production of keratin results in a hyperkeratotic type of BCC. There can also be sebaceous and adenoid differentiation while the growth of elongated strands and islands of cells embedded in a dense fibrous stroma results in a sclerosing (morphoeic) type of tumour.

Clinical features

Eyelid BCC generally conforms to one of the morphological patterns below.

- **Nodular BCC** is a shiny, firm, pearly nodule with small overlying dilated blood vessels. Initially, growth is slow and it may take the tumour 1–2 years to reach a diameter of 0.5 cm (Figs 1.19A and B).
- **Noduloulcerative BCC (rodent ulcer)** is centrally ulcerated with pearly raised rolled edges and dilated and irregular blood vessels (telangiectasis) over its lateral margins (Fig. 1.19C); with time it may erode a large portion of the eyelid (Fig. 1.19D).
- **Sclerosing (morphoeic) BCC** is less common and may be difficult to diagnose because it infiltrates laterally beneath

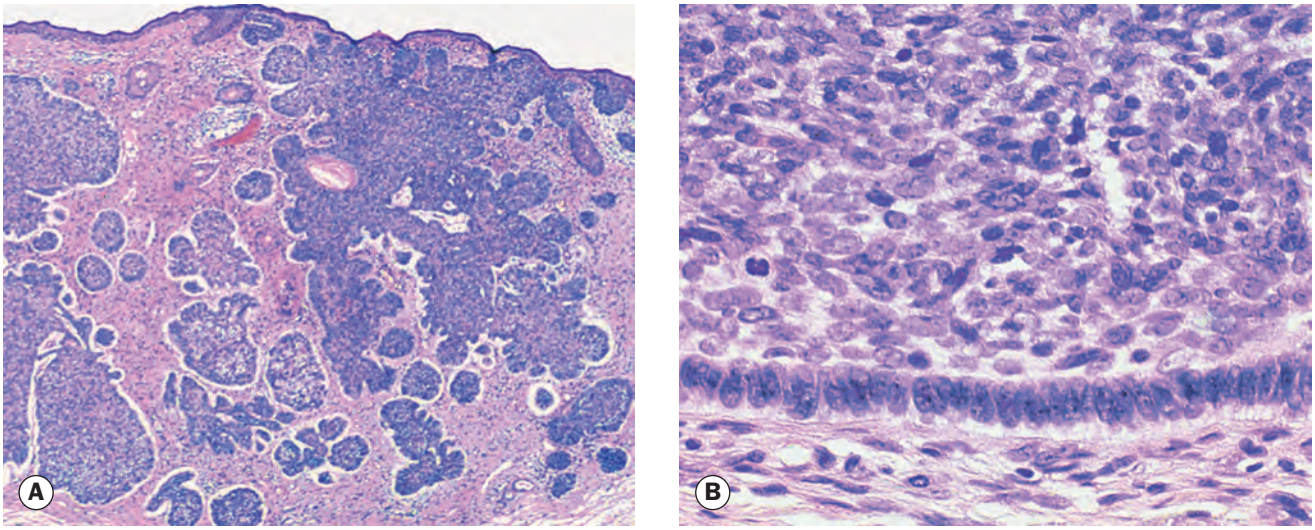


Fig. 1.18 Histopathology of basal cell carcinoma. **(A)** Histopathology shows downward proliferation of lobules of basophilic (purple) cells; **(B)** palisading of cells at the periphery of a tumour lobule
(Courtesy of J Harry)

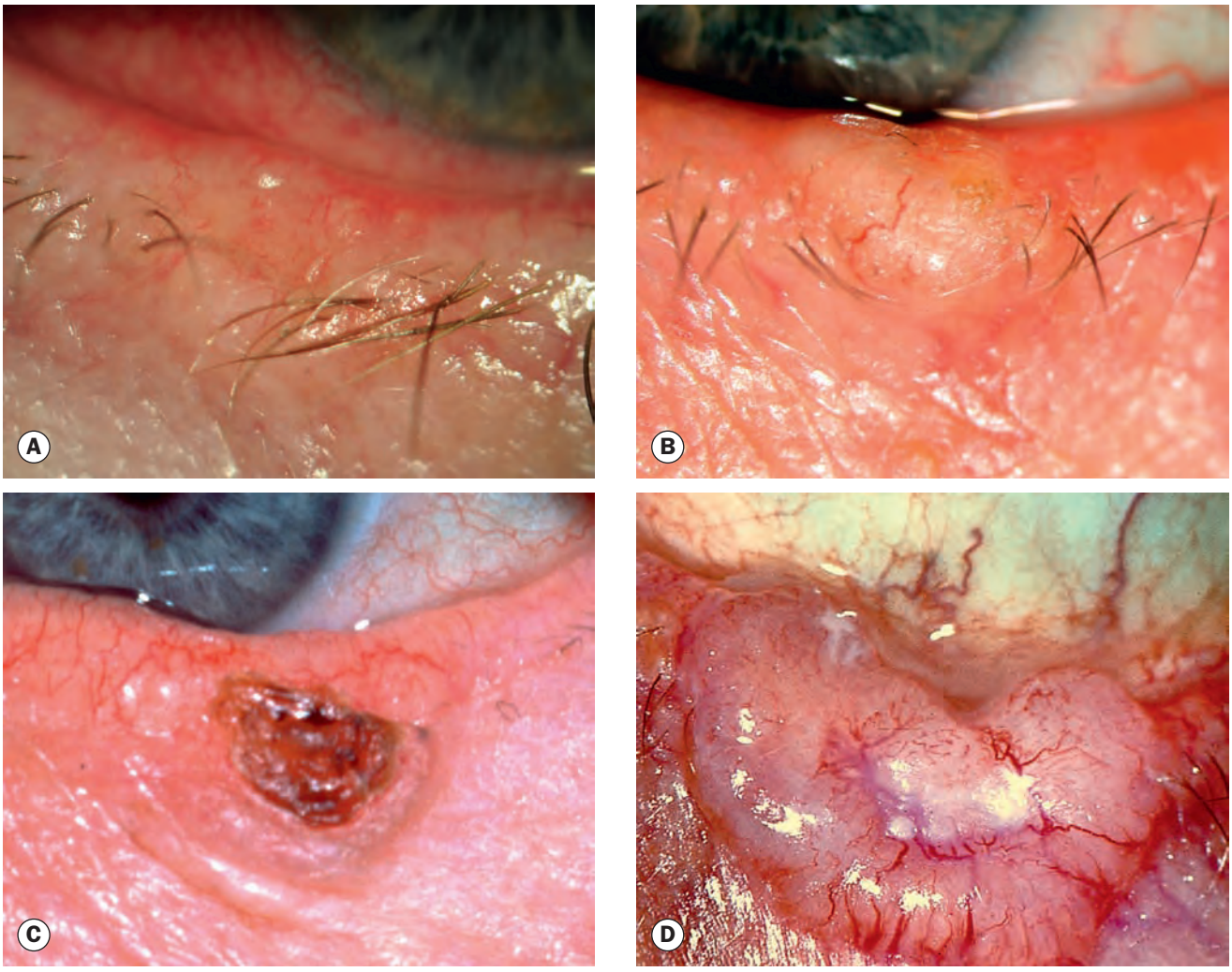


Fig. 1.19 Clinical appearance of basal cell carcinoma. **(A)** Early nodular lesion; **(B)** larger nodular tumour; **(C)** rodent ulcer; **(D)** large rodent ulcer;

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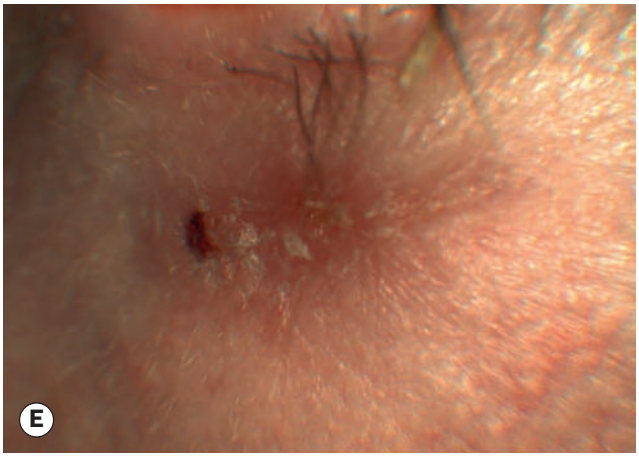


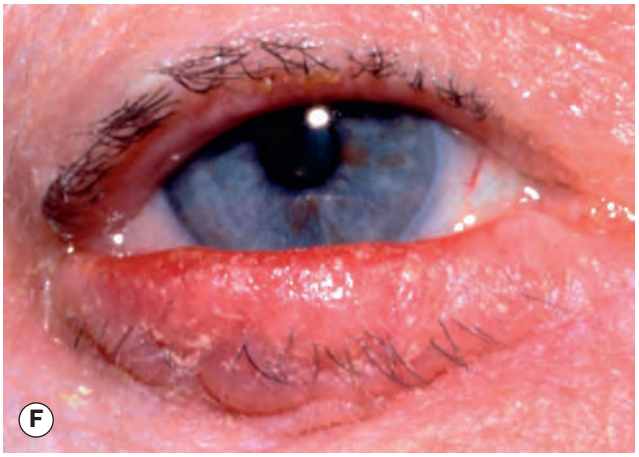


Fig. 1.19, Continued **(E)** sclerosing tumour; **(F)** extensive sclerosing tumour

the epidermis as an indurated plaque (Figs 1.19E and F). The margins of the tumour may be impossible to delineate clinically and the lesion tends to be much more extensive on palpation than inspection. On cursory examination a sclerosing BCC may simulate a localized area of chronic blepharitis.

- **Other types** not usually found on the lid are cystic, adenoid, pigmented and multiple superficial.

Squamous cell carcinoma

Introduction

SCC is a much less common, but typically more aggressive tumour than BCC with metastasis to regional lymph nodes in about 20% of cases. Careful surveillance of regional lymph nodes is therefore an important aspect of initial management. The tumour may also exhibit perineural spread to the intracranial cavity via the orbit. SCC accounts for 5–10% of eyelid malignancies and may arise *de novo* or from pre-existing actinic keratosis or carcinoma *in situ* (Bowen disease, intraepidermal carcinoma – Fig. 1.20). Immunocompromised patients, such as those with acquired immunodeficiency syndrome (AIDS) or following renal transplantation are at increased risk, as are those with a predisposing syndrome such as xeroderma pigmentosum. The tumour has a predilection for the lower eyelid and the lid margin. It occurs most commonly in older individuals with a fair complexion and a history of chronic sun exposure. The diagnosis of SCC may be difficult because certain ostensibly benign lesions such as keratoacanthoma and cutaneous horn may reveal histological evidence of invasive SCC at deeper levels of sectioning.

Histopathology

The tumour arises from the squamous cell layer of the epidermis. It is composed of variably sized groups of atypical epithelial cells with prominent nuclei and abundant eosinophilic cytoplasm within the dermis (Fig. 1.21A). Well-differentiated tumours may show characteristic keratin 'pearls' and intercellular bridges (desmosomes).

Clinical features

The clinical types are variable and there are no pathognomonic characteristics. The tumour may be indistinguishable clinically from a BCC but surface vascularization is usually absent, growth is more rapid and hyperkeratosis is more common.

- **Nodular SCC** is characterized by a hyperkeratotic nodule that may develop crusting, erosions and fissures (Fig. 1.21B).
- **Ulcerating SCC** has a red base and sharply defined, indurated and everted borders, but pearly margins and telangiectasia are not usually present (Fig. 1.21C).
- **Cutaneous horn** with underlying invasive SCC (Fig. 1.21D).

Fig. 1.20 Carcinoma *in situ*
(Courtesy of H Frank)

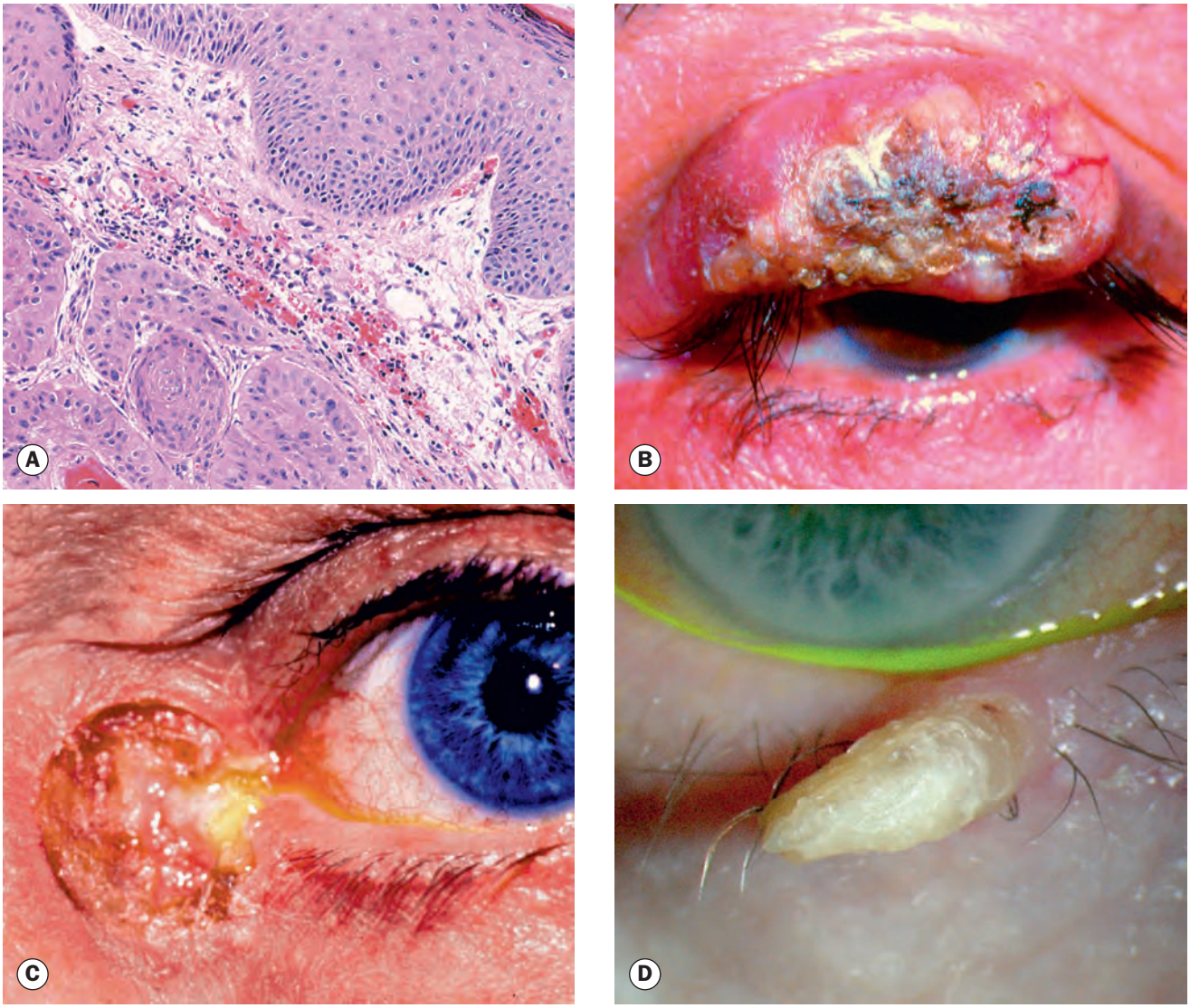


Fig. 1.21 Squamous cell carcinoma. **(A)** Histopathology shows acanthotic squamous epithelium and eosinophilic (pink) islands of dysplastic squamous epithelium within the dermis; **(B)** nodular tumour with surface keratosis; **(C)** ulcerating tumour; **(D)** cutaneous horn

(Courtesy of L Horton – fig. A; A Singh, from Clinical Ophthalmic Oncology, *Saunders 2007 – fig. B; H Frank – fig. C; S Farley, T Cole and L Rimmer – fig. D)*

Keratoacanthoma

Introduction

Keratoacanthoma is a rare, rapidly growing but subsequently regressing tumour that usually occurs in fair-skinned individuals with a history of chronic sun exposure. Immunosuppressive therapy is also a predisposing factor. It is regarded as falling within the spectrum of SCC, and although invasion and metastasis are rare, definitive treatment is usually indicated. Histopathologically, irregular thickened epidermis is surrounded by acanthotic squamous epithelium; a sharp transition from the thickened involved area to normal adjacent epidermis is referred to as shoulder formation (Fig. 1.22A); a keratin-filled crater may be seen.

Diagnosis

A pink dome-shaped hyperkeratotic lesion develops, often on the lower lid (Fig. 1.22B), and may double or treble in size within weeks (Fig. 1.22C). Growth then ceases for 2–3 months, after which spontaneous involution occurs, when a keratin-filled crater may develop (Fig. 1.22D). Complete involution may take up to a year and usually leaves an unsightly scar.

Treatment

Treatment generally involves complete surgical excision with a margin of at least 3 mm, or utilizing Mohs surgery; radiotherapy, cryotherapy or local chemotherapy are sometimes used. Observation is now regarded as inappropriate.

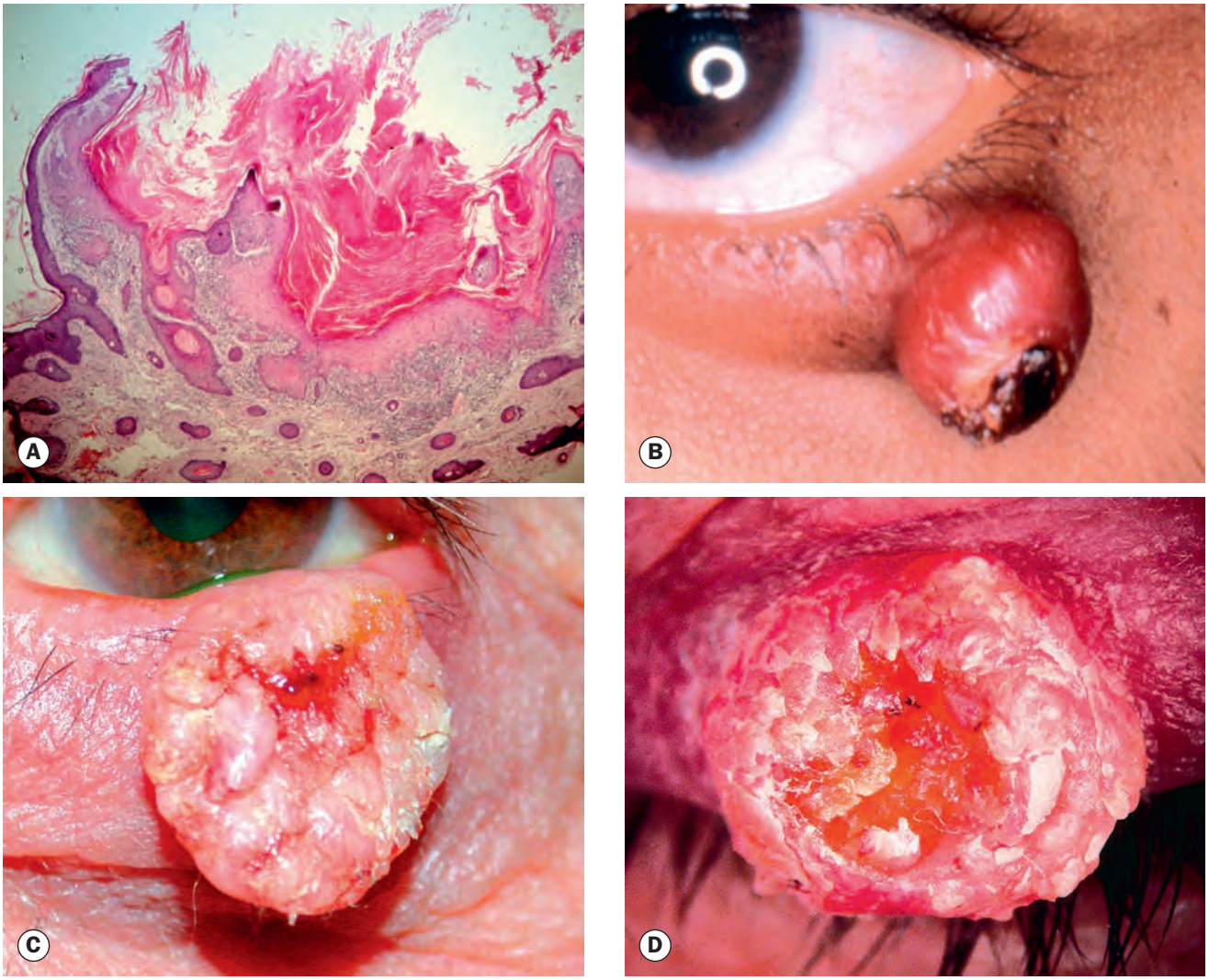


Fig. 1.22 Keratoacanthoma. **(A)** Histopathology shows irregularly thickened eosinophilic epidermis with a keratin-containing cup and well-marked shoulder formation; **(B)** hyperkeratotic nodule; **(C)** large tumour; **(D)** keratin-filled crater during involution

Sebaceous gland carcinoma

Introduction

Sebaceous gland carcinoma (SGC) is a very rare, slowly growing tumour that most frequently affects the elderly, with a predisposition for females. It usually arises from the meibomian glands, although on occasion it may arise from the glands of Zeis or elsewhere. The tumour consists histopathologically of lobules of cells with pale foamy vacuolated lipid-containing cytoplasm and large hyperchromatic nuclei (Fig. 1.23A). Pagetoid spread refers to extension of a tumour within the epithelium, and is not uncommon. Overall mortality is 5–10%; adverse prognostic features include upper lid involvement, tumour size of 10 mm or more and duration of symptoms of more than 6 months.

Clinical features

In contrast to BCC and SCC, SGC occurs more commonly on the upper eyelid where meibomian glands are more numerous; there may be simultaneous involvement of both lids on one side (5%).

- **Yellowish material** within the tumour is highly suggestive of SGC.
- **Nodular SGC** presents as a discrete, hard nodule, most commonly within the upper tarsal plate (Fig. 1.23B), and may exhibit yellow discoloration due to the presence of lipid; it can be mistaken for a chalazion.
- **Spreading SGC** infiltrates into the dermis and causes a diffuse thickening of the lid margin (Fig. 1.23C) often with eyelash distortion and loss, and can be mistaken for blepharitis.

Lentigo maligna and melanoma

Introduction

Melanoma rarely develops on the eyelids but is potentially lethal. Although pigmentation is a hallmark of skin melanomas, half of

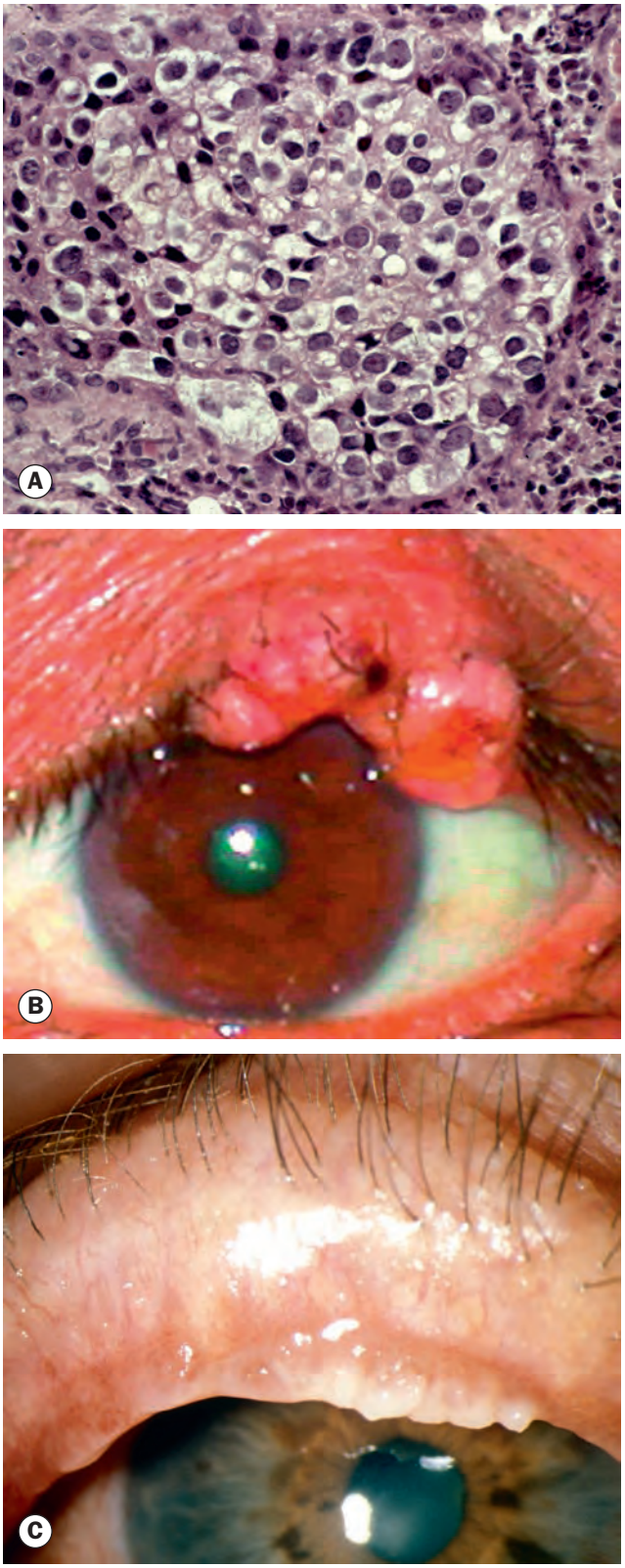


Fig. 1.23 Sebaceous gland carcinoma. **(A)** Histopathology shows cells with large hyperchromatic nuclei and vacuolated cytoplasm; **(B)** nodular tumour; **(C)** spreading tumour

(Courtesy of A Garner – fig. A; A Singh, from Clinical Ophthalmic Oncology, *Saunders 2007 – fig. B; S Tuft – fig. C)*

lid melanomas are non-pigmented and this may give rise to diagnostic difficulty. Features suggestive of melanoma include recent onset of a pigmented lesion, change in an existing pigmented lesion, irregular margins, asymmetrical shape, colour change or presence of multiple colours, and diameter greater than 6 mm.

Lentigo maligna

Lentigo maligna (melanoma *in situ*, intraepidermal melanoma, Hutchinson freckle) is an uncommon condition that develops in sun-damaged skin in elderly individuals. Malignant change may occur, with infiltration of the dermis. Histopathology shows intraepidermal proliferation of spindle-shaped atypical melanocytes replacing the basal layer of the epidermis (Fig. 1.24A). Clinically lentigo maligna presents as a slowly expanding pigmented macule with an irregular border (Fig. 1.24B). Treatment is usually by excision. Nodular thickening and areas of irregular pigmentation are highly suggestive of malignant transformation (Fig. 1.24C).

Melanoma

Histopathology shows large atypical melanocytes invading the dermis (Fig. 1.25A). Superficial spreading melanoma is characterized by a plaque with an irregular outline and variable pigmentation (Fig. 1.25B). Nodular melanoma is typically a blue – black nodule surrounded by normal skin (Fig. 1.25C). Treatment is usually by wide excision and may include local lymph node removal. Radiotherapy, chemotherapy, biological and 'targeted' therapy may also be used, generally as adjuvants.

Merkel cell carcinoma

Merkel cells are a form of sensory receptor concerned with light touch. Merkel cell carcinoma is a rapidly growing, highly malignant tumour that typically affects older adults. Its rarity may lead to difficulty in diagnosis and delay in treatment, and 50% of patients have metastatic spread by presentation. A violaceous, well-demarcated nodule with intact overlying skin is seen, most frequently involving the upper eyelid (Fig. 1.26). Treatment is by excision, often with adjuvant therapy.

Kaposi sarcoma

Kaposi sarcoma is a vascular tumour that typically affects patients with AIDS. Many patients have advanced systemic disease although in some instances the tumour may be the only clinical manifestation of human immunodeficiency virus (HIV) infection. Histopathology shows proliferating spindle cells, vascular channels and inflammatory cells within the dermis (Fig. 1.27A). Clinically a pink, red-violet to brown lesion (Fig. 1.27B) develops, which may be mistaken for a haematoma or naevus. Treatment is by radiotherapy or excision, and by optimal control of AIDS where relevant.

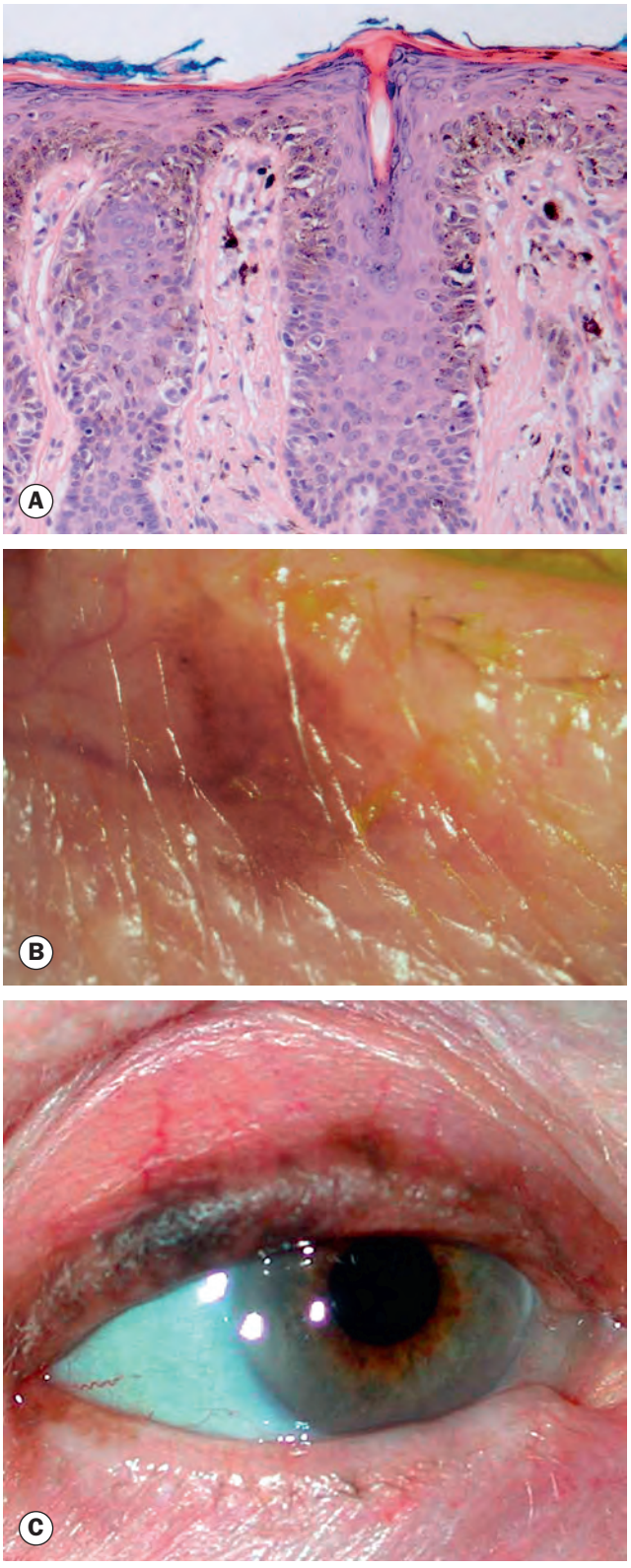


Fig. 1.24 Lentigo maligna of the eyelid. **(A)** Histopathology shows melanoma cells proliferating within the basal layers of the epidermis; **(B)** early lentigo maligna; **(C)** melanoma arising from lentigo maligna

(Courtesy of L Horton – fig. A; S Delva – fig. C)

Fig. 1.25 Melanoma. **(A)** Histopathology shows melanoma cells within the dermis; **(B)** superficial spreading melanoma; **(C)** nodular melanoma

(Courtesy of J Harry – fig. A)

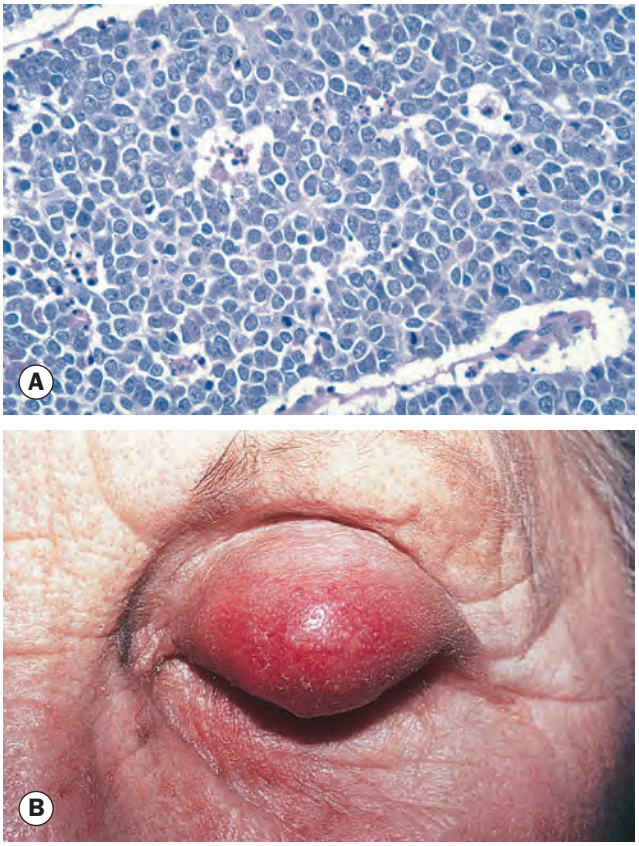


Fig. 1.26 Merkel cell carcinoma. **(A)** Histopathology shows a sheet of Merkel cells; **(B)** clinical appearance

(Courtesy of J Harry and G Misson, from Clinical Ophthalmic Pathology, *Butterworth-Heinemann 2001 – fig. A)*

Treatment of malignant tumours

Biopsy

Biopsy can be (i) *incisional*, using a blade or a biopsy punch, in which only part of the lesion is removed for histological diagnosis, or (ii) *excisional*, in which the entire lesion is removed; the latter may consist of shave excision using a blade to remove shallow epithelial tumours, such as papillomas and seborrhoeic keratosis, or full-thickness skin excision for tumours that are not confined to the epidermis.

Surgical excision

Surgical excision aims to remove the entire tumour with preservation of as much normal tissue as possible. Smaller tumours can be removed via an excision biopsy and the defect closed directly, whilst awaiting histological confirmation of complete clearance. Most small BCCs can be cured by excision of the tumour together with a 2–4 mm margin of clinically normal tissue. More radical surgical excision is required for large BCCs and aggressive tumours such as SCC, SGC and melanoma. It may not be possible to close all defects at the time of initial removal, but it is necessary to ensure complete clearance of tumour prior to undertaking any reconstruction. There are several options for the coordination of histopathological diagnosis and tumour clearance with excision.

- **Conventional paraffin-embedded specimen.** Rapid processing can reduce the interval to confirmation of histological clearance but still requires that reconstruction be performed as a separate procedure. Faster confirmation can be achieved using either frozen-section control or micrographic surgery (see next), and reconstruction can then take place on the same day.
- **Standard frozen section** involves histological examination of the margins of the excised specimen at the time of surgery to ensure they are tumour-free. If no tumour cells are detected, the eyelid is reconstructed on the same day; if residual tumour is present, further excision is performed at the appropriate edge of the surgical site until no tumour is detected.
- **Mohs micrographic surgery** involves layered excision of the tumour; specimens are usually examined frozen. Processing of each layer enables a map of the edges of the tumour to be

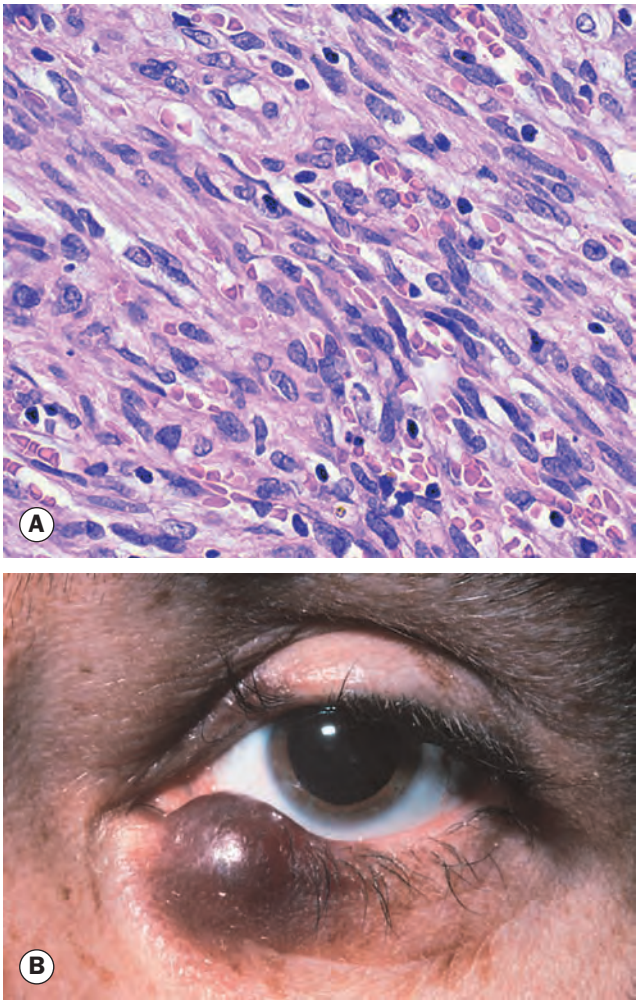


Fig. 1.27 Kaposi sarcoma. **(A)** Histopathology shows a proliferation of predominantly spindle-shaped cells; vascular channels are evident; **(B)** clinical appearance

(Courtesy of J Harry – fig. A)

developed. Further tissue is taken in any area where tumour is still present until clearance is achieved. Although time-consuming, this technique maximizes the chances of total tumour excision whilst minimizing sacrifice of normal tissue. This is a particularly useful technique for tumours that grow diffusely and have indefinite margins with finger-like extensions, such as sclerosing BCC, SCC, recurrent tumours and those involving the medial or lateral canthi. The irregular contours around the eyelids and extension of tumours into orbital fat can make interpretation difficult.

Reconstruction

The technique of reconstruction depends on the extent of tissue removed. It is important to reconstruct both anterior and posterior lamellae, each of which must be reconstructed with similar tissue. Anterior lamellar defects may be closed directly or with a local flap or skin graft. Options for the repair of full-thickness defects are set out below.

- **Small defects** involving less than one-third of the eyelid can usually be closed directly, provided the surrounding tissue is sufficiently elastic to allow approximation of the cut edges (Fig. 1.28). If necessary, a lateral cantholysis can be performed for increased mobilization.
- **Moderate size defects** involving up to half of the eyelid may require a flap (e.g. Tenzel semicircular) for closure (Fig. 1.29).
- **Large defects** involving over half of the eyelid may be closed by one of the following techniques:
  - Posterior lamellar reconstruction may involve an upper lid free tarsal graft, buccal mucous membrane or hard palate graft, or a Hughes tarsoconjunctival flap from the upper lid, which is left attached for 4–6 weeks before transection (Fig. 1.30).
  - Anterior lamellar reconstruction may involve skin advancement, a local skin flap or a free skin graft (Fig. 1.31); the patient must be made aware that grafted skin is unlikely to be a perfect match. At least one reconstructed lamella requires its own blood supply to maximize the viability of a free graft component.

Laissez-faire

Full reconstruction of the defect created by tumour removal may not always be required. In the *laissez-faire* approach the wound edges are approximated as far as possible and the defect is allowed to granulate and heal by secondary intention. Even large defects can often achieve a satisfactory outcome with time.

Radiotherapy

The recurrence rate following irradiation alone is higher than after surgery, and radiotherapy does not allow histological confirmation of tumour eradication. Recurrences following radiotherapy are difficult to treat surgically because of the poor healing properties of irradiated tissue. However, it still has utility in some circumstances.

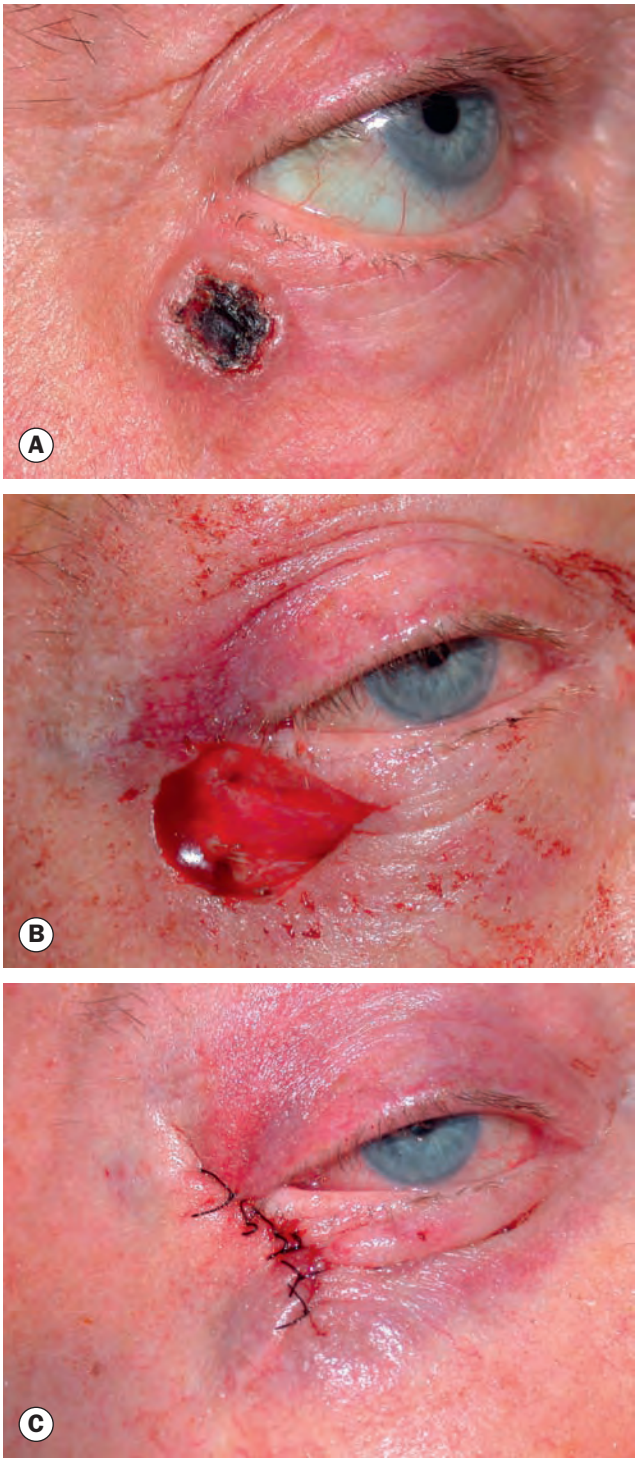
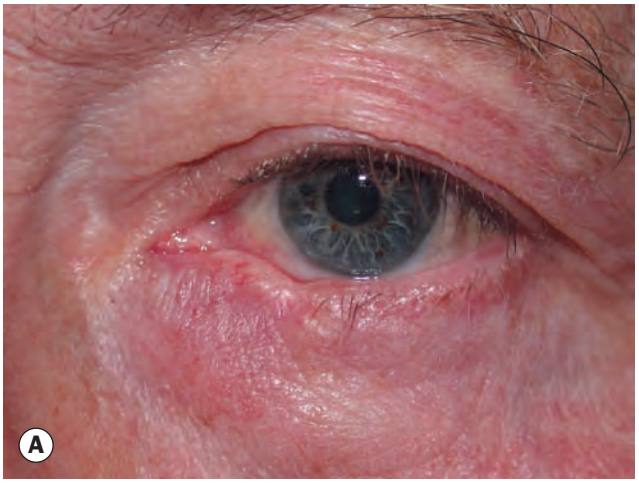


Fig. 1.28 Direct closure. **(A)** Preoperative appearance of a basal cell carcinoma; **(B)** appearance following excision; **(C)** direct closure of defect
(Courtesy of A Pearson)

- **Indications**
  - Patients who are either unsuitable for or refuse surgery.
  - Highly radiosensitive tumours, such as Kaposi sarcoma.
  - Adjunctive therapy in some cases.
  - Palliative treatment.



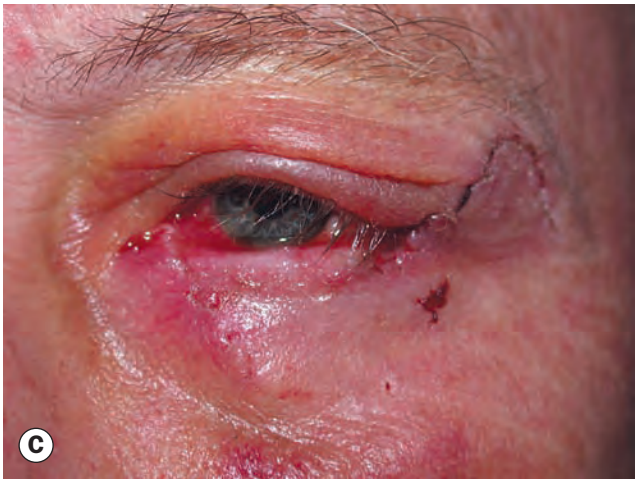


Fig. 1.29 Tenzel flap. **(A)** Preoperative appearance; **(B)** appearance following excision; **(C)** appearance following closure of the flap

(Courtesy of A Pearson)

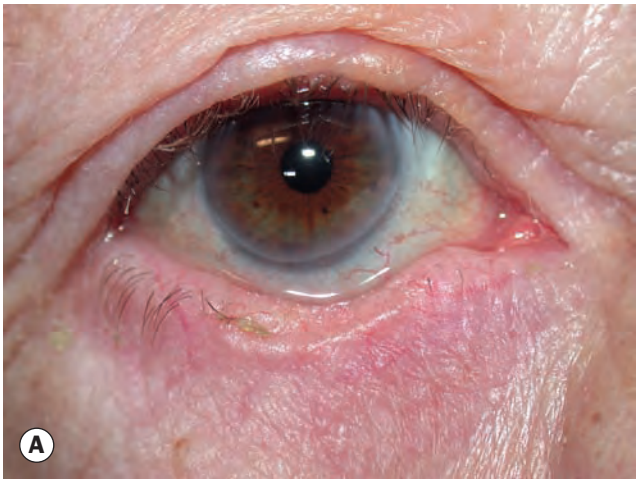


Fig. 1.30 Posterior lamellar reconstruction with a Hughes upper lid flap. **(A)** Preoperative appearance; **(B)** appearance following excision; **(C)** postoperative appearance with the flap yet to be divided

(Courtesy of A Pearson)

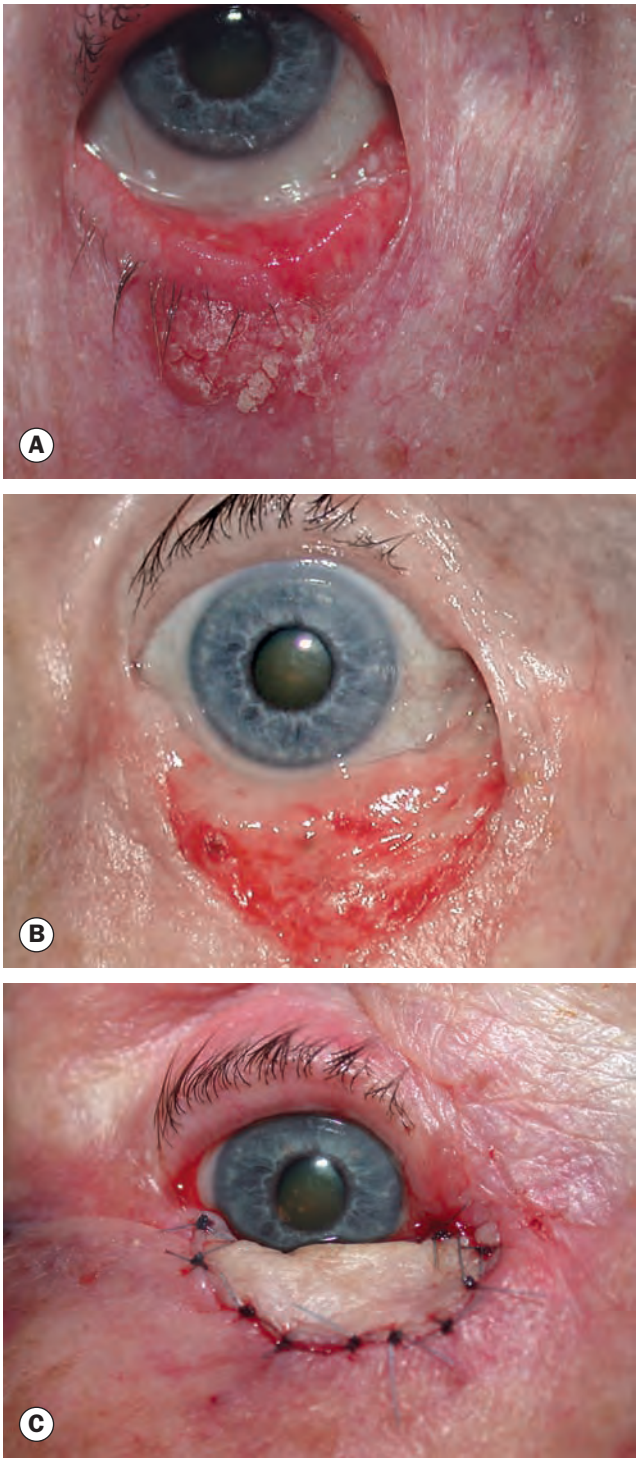


Fig. 1.31 Anterior lamellar reconstruction with a free skin graft. **(A)** Preoperative appearance; **(B)** appearance following excision; **(C)** skin graft in place
(Courtesy of A Pearson)

- **Relative contraindications**
  - Medial canthal lesions due to the high probability of lacrimal canalicular damage.
  - Upper eyelid tumours – conjunctival keratinization is common and difficult to manage.
  - Aggressive tumours such as SGC are relatively radioresistant, but higher-dose treatment may be effective.
- **Complications.** Many of these can be minimized by appropriate shielding.
  - Skin damage and madarosis (eyelash loss).
  - Nasolacrimal duct stenosis following irradiation to the medial canthal area.
  - Conjunctival keratinization, dry eye, keratopathy and cataract.
  - Retinopathy and optic neuropathy.

Cryotherapy

Cryotherapy may be considered for small superficial BCCs; it can be a useful adjunct to surgery in some patients. Complications include skin depigmentation, madarosis and conjunctival overgrowth.

DISORDERS OF THE EYELASHES

Misdirected lashes

Introduction

The roots of the eyelashes (cilia) lie against the anterior surface of the tarsal plate. The cilia pass between the main part of the orbicularis oculi and its more superficial part (Riolan muscle), exiting the skin at the anterior lid margin and curving away from the globe. It is particularly important to be familiar with the normal anatomical appearance of the lid margin in order to be able to identify the cause of eyelash misdirection. From anterior to posterior:

- **Eyelashes** (cilia).
- **The grey line**, by definition the border between the anterior (lashes, skin and orbicularis) and posterior (tarsal plate and conjunctiva) lamellae.
- **The meibomian gland orifices** are located just anterior to the mucocutaneous junction. The edge of the tarsal plate is deep to the gland orifices; the glands themselves run vertically within the plate.
- **The mucocutaneous junction** is where keratinized epithelium of the skin merges with conjunctival mucous membrane.
- **Conjunctiva** lines the posterior margin of the lid.

Clinical features

Trauma to the corneal epithelium may cause punctate epithelial erosions, with ocular irritation often worsened by blinking. Corneal ulceration and pannus formation may occur in severe cases. The clinical appearance varies with the cause.

- **Trichiasis** refers to misdirection of growth from individual follicles (Fig. 1.32A), rather than a more extensive inversion of the lid or lid margin. The follicles are at anatomically normal sites. It is commonly due to inflammation such as chronic blepharitis or herpes zoster ophthalmicus, but can

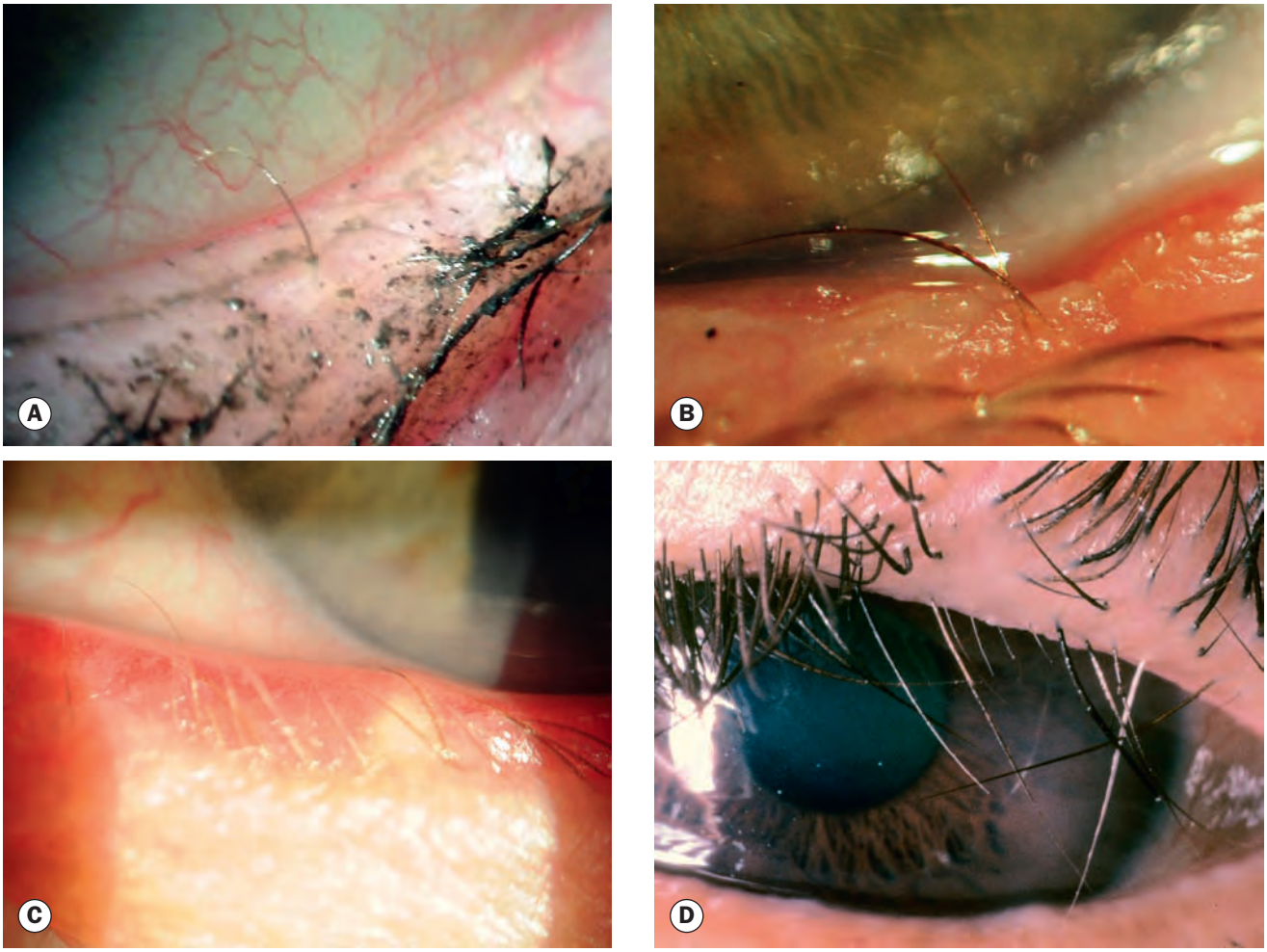


Fig. 1.32 Misdirected lashes. **(A)** Single trichiatic lash; **(B)** trichiasis associated with a lid notch following chalazion surgery; **(C)** marginal entropion showing rows of misdirected lashes, anterior migration of the mucocutaneous junction, and a rounded posterior lid margin; **(D)** acquired distichiasis
(Courtesy of S Chen – fig. A; R Bates – fig. D)

also be caused by trauma, including surgery such as incision and curettage of a chalazion (Fig. 1.32B).

- **Marginal entropion** has increasingly been recognized as a very common cause of eyelash misdirection, the mechanism of which is thought to be subtle cicatricial posterior lamellar shortening that rotates a segment of the lid margin towards the eye. The mucocutaneous junction migrates anteriorly and the posterior lid margin becomes rounded rather than physiologically square. Typically, numerous aligned lashes are involved (Fig. 1.32C).
- **Congenital distichiasis** is a rare condition that occurs when a primary epithelial germ cell destined to differentiate into a meibomian gland develops instead into a complete pilosebaceous unit. The condition is frequently inherited in an autosomal dominant manner with high penetrance but variable expressivity. The majority of patients also manifest primary lymphoedema of the legs (lymphoedema–distichiasis syndrome). A partial or complete second row of lashes is seen to emerge at or slightly behind the meibomian gland orifices. The aberrant lashes tend to be thinner and shorter than normal cilia and are often directed posteriorly. They are usually well tolerated during infancy and may not become symptomatic until the age of about 5 years.
- **Acquired distichiasis** is caused by metaplasia of the meibomian glands into hair follicles such that a variable number of lashes grow from meibomian gland openings. The most important cause is intense conjunctival inflammation (e.g. chemical injury, Stevens–Johnson syndrome, ocular cicatricial pemphigoid). In contrast to congenital distichiasis, the cilia tend to be non-pigmented and stunted (Fig. 1.32D), and are usually symptomatic.
- **Epiblepharon** – see later.
- **Entropion.** In contrast to marginal entropion, profound inversion of a substantial width of the lid is readily identified – see later.

Treatment

- **Epilation** with forceps is simple and effective but recurrence within a few weeks is essentially invariable. It can be used as

a temporizing measure or in the occasional patient who refuses or cannot tolerate surgery.

- **Electrolysis** or electrocautery (hyfrecation) are broadly similar electrosurgical techniques in which, under local anaesthesia, a fine wire is passed down the hair follicle to ablate the lash. It is generally useful for a limited number of lashes; scarring can occur. Frequently multiple treatments are required to obtain a satisfactory result.
- **Laser ablation** is also useful for the treatment of limited aberrant eyelashes, and is performed using a spot size of 50 µm, duration of 0.1–0.2 s and power of 800–1000 mW. The base of the lash is targeted and shots are applied to create a crater that follows the axis of the follicle (Fig. 1.33). Success is broadly comparable to that achieved with electrosurgery.
- **Surgery**
  - Tarsal facture (transverse tarsotomy) is performed for marginal entropion. After placing a 4-0 traction suture, a horizontal incision is made through the tarsal plate via the conjunctiva, at least halfway down the plate, along the affected length of the lid and extended to 2–3 mm either side of the involved region. Depending on the extent of lid involvement, either two or three double-armed absorbable sutures are passed through the upper edge of the lower section of the tarsal plate to emerge just anterior to the lashes, leaving the lid margin very slightly everted (Fig. 1.34). The sutures are left in place following the surgery; occasionally short-term use of a bandage contact lens is required to prevent corneal abrading.
  - A full-thickness eyelid pentagon resection can be used for a focal group of aberrant lashes, typically after trauma, or for localized marginal entropion.
  - Other options include lid splitting (see next) with follicle excision, and anterior lamellar rotation surgery.
- **Cryotherapy** applied externally to the skin just inferior to the base of the abnormal lashes or – especially in distichiasis – to the internal aspect of the anterior lamella of the lid following splitting of the margin at the grey line (Fig. 1.35), can be used for numerous lashes. A double freeze–thaw cycle at −20 °C is applied under local anaesthesia (including adrenaline) with a plastic eye protector in place; suturing of the lid margin is not usually necessary following limited splitting. The method is effective but carries a high rate of local adverse effects, and is less commonly performed than previously.

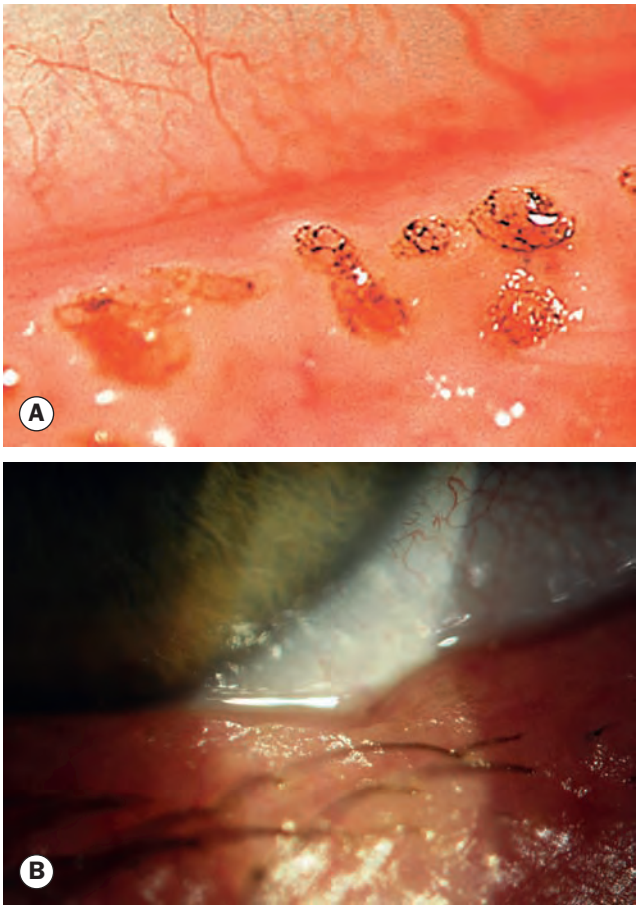


Fig. 1.33 Laser for trichiasis. **(A)** Appearance following ablation of multiple lashes; **(B)** the eye in Fig. 1.32B 6 weeks after laser ablation

Eyelash ptosis

Eyelash ptosis refers to a downward sagging of the upper lid lashes (Fig. 1.36A). The condition may be idiopathic or associated with floppy eyelid syndrome, dermatochalasis with anterior lamellar slip or long-standing facial palsy.

Trichomegaly

Trichomegaly is excessive eyelash growth (Fig. 1.36B); the main causes are listed in Table 1.1.

Madarosis

Madarosis is the term used for the loss of lashes (Fig. 1.36C). The main causes are shown in Table 1.2.

Poliosis

Poliosis is a premature localized whitening of hair, which may involve the lashes and eyebrows (Fig. 1.36D); the main causes are shown in Table 1.3.

Table 1.1 Causes of trichomegaly

Drug-induced – topical prostaglandin analogues, phenytoin and ciclosporin
Malnutrition
AIDS
Porphyria
Hypothyroidism
Familial
Congenital: Oliver–McFarlane, Cornelia de Lange, Goldstein–Hutt, Hermansky–Pudlak syndromes

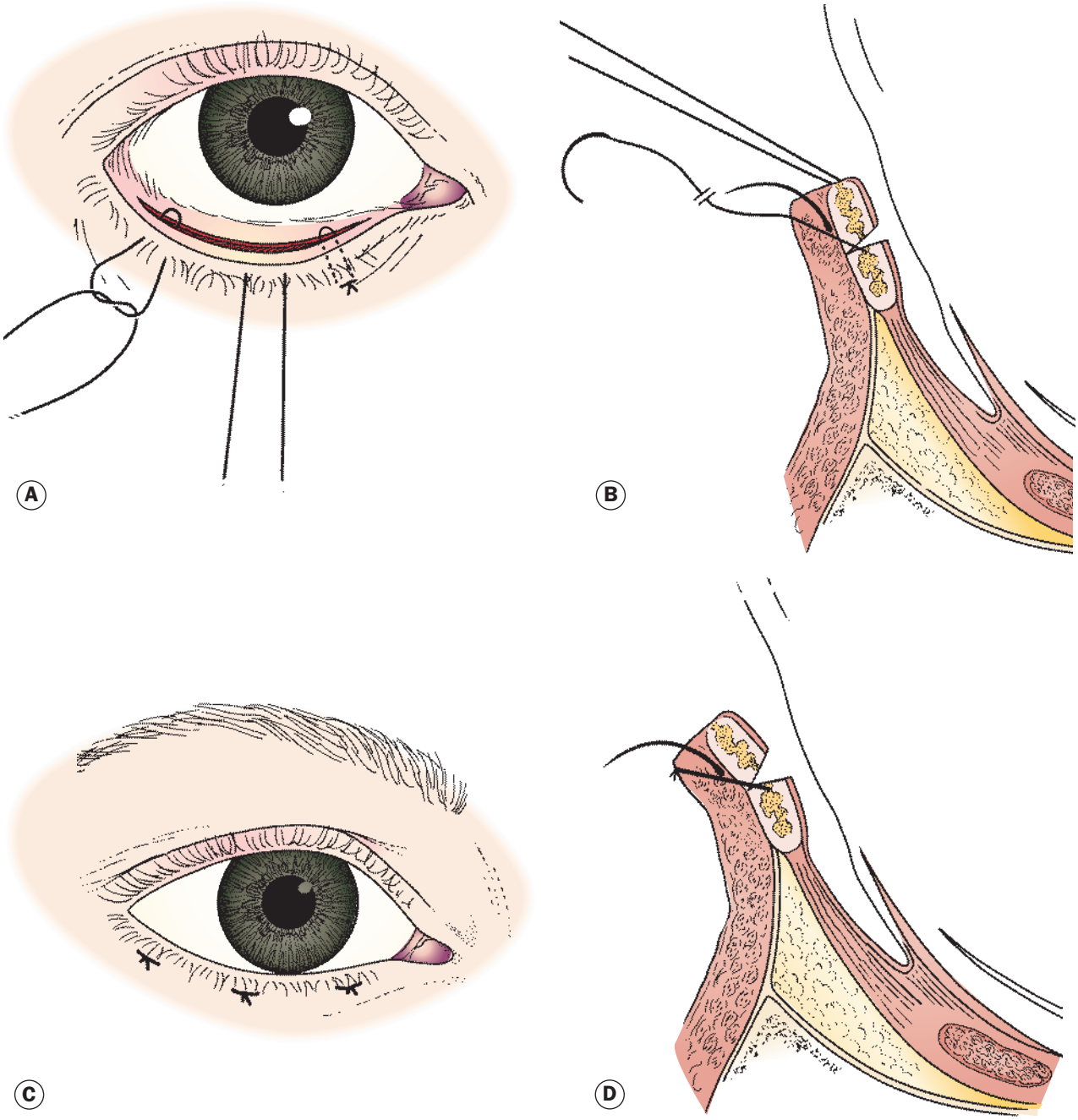


Fig. 1.34 Tarsal fracture for repair of marginal entropion. **(A)** and **(B)** insertion of everting sutures following traction suture emplacement and horizontal tarsal plate incision; **(C)** and **(D)** everting sutures in place
(Courtesy of JA Nerad, from Techniques in Ophthalmic Plastic Surgery, *Saunders 2010)*

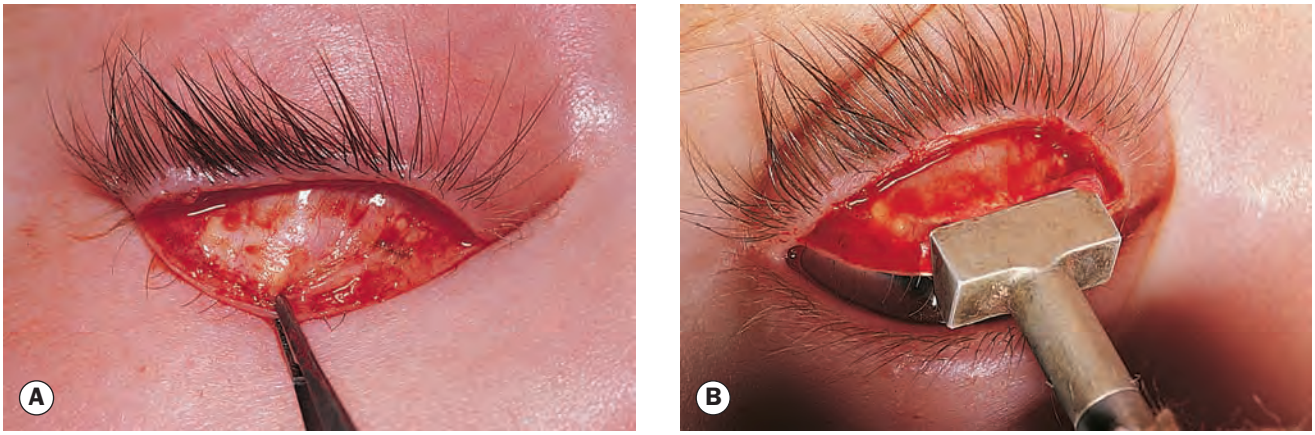


Fig. 1.35 Cryotherapy to the eyelid in distichiasis. **(A)** Separation of the anterior and posterior lamellae; **(B)** application of cryoprobe to the posterior lamella

(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001)*

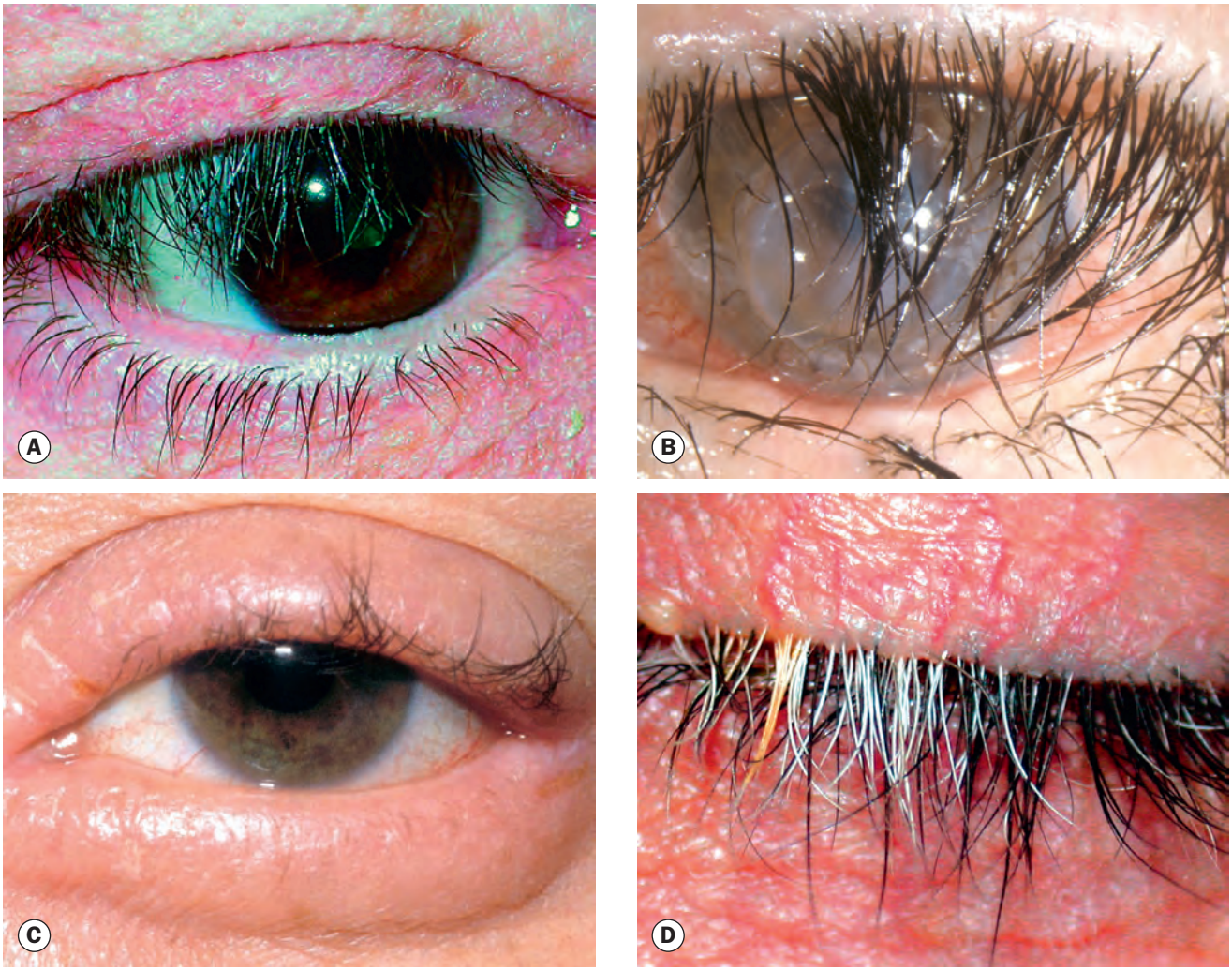


Fig. 1.36 Miscellaneous eyelash disorders. **(A)** Eyelash ptosis; **(B)** trichomegaly; **(C)** madarosis; **(D)** poliosis

(Courtesy of A Pearson – fig. A; L Merin – fig. B; S Tuft – fig. C)

Table 1.2 Cause of madarosis

1. Local
Chronic anterior lid margin disease
Infiltrating lid tumours
Burns
Radiotherapy or cryotherapy of lid tumours
2. Skin disorders
Generalized alopecia
Psoriasis
3. Systemic diseases
Myxoedema
Systemic lupus erythematosus
Acquired syphilis
Lepromatous leprosy
4. Following removal
Procedures for trichiasis
Trichotillomania – psychiatric disorder of hair removal

Table 1.3 Causes of poliosis

1. Ocular
Chronic anterior blepharitis
Sympathetic ophthalmitis
Idiopathic uveitis
2. Systemic
Vogt–Koyanagi–Harada syndrome
Waardenburg syndrome
Vitiligo
Marfan syndrome
Tuberous sclerosis

ALLERGIC DISORDERS

Acute allergic oedema

Acute allergic oedema is usually caused by exposure to pollen or by insect bites, and manifests with the sudden onset of bilateral boggy periocular oedema (Fig. 1.37A), often accompanied by conjunctival swelling (chemosis – see Ch. 5). Treatment is often unnecessary, but systemic antihistamines are sometimes given.

Contact dermatitis

Contact dermatitis is an inflammatory response that usually follows exposure to a medication such as eye drops (often preservative-containing), cosmetics or metals. An irritant can also cause a non-allergic toxic dermatitis. The individual is sensitized on first exposure and develops an immune reaction on further exposure; the mediating reaction is type IV (delayed type) hypersensitivity. Signs consist of lid skin scaling, angular fissuring, oedema and tightness (Fig. 1.37B); there may be chemosis, redness and papillary conjunctivitis. Corneal involvement is usually limited to punctate epithelial erosions. Treatment consists primarily of avoidance of allergen exposure, provided it can be identified. Cold compresses provide symptomatic relief. Topical steroids and oral antihistamines can be used, but are rarely required.

Atopic dermatitis

Atopic dermatitis (eczema) is a very common idiopathic condition, typically occurring in patients who also suffer from asthma and hay fever. Eyelid involvement is relatively infrequent but when present is invariably associated with generalized dermatitis. Thickening, crusting and fissuring of the lids (Fig. 1.37C) is typical, and staphylococcal blepharitis, vernal or atopic keratoconjunctivitis are also commonly present. Herpetic blepharitis and keratoconjunctivitis is more common and more severe in patients with atopy (eczema herpeticum). Treatment of the lid features is with emollients to hydrate the skin and the judicious use of mild topical steroid such as hydrocortisone 1%. Uncommon ocular associations include keratoconus, cataract and retinal detachment (see also Ch. 5).

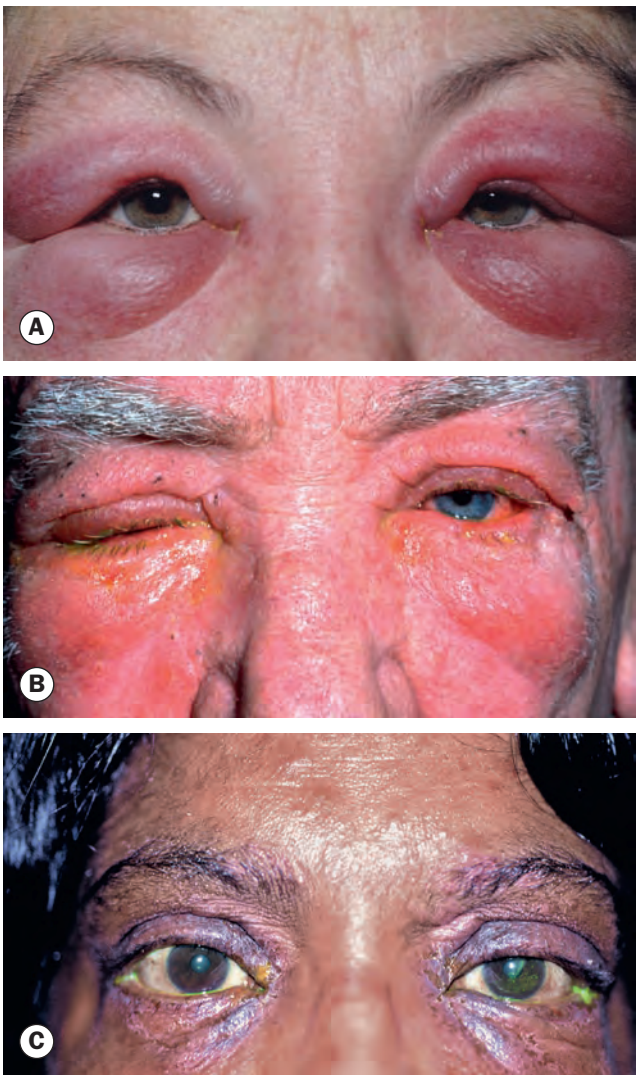


Fig. 1.37 Allergic disorders. **(A)** Acute allergic oedema; **(B)** contact dermatitis; **(C)** atopic dermatitis

BACTERIAL INFECTIONS

External hordeolum

An external hordeolum (stye) is an acute staphylococcal abscess of a lash follicle and its associated gland of Zeis that is common in children and young adults. A stye presents as a tender swelling in the lid margin pointing anteriorly through the skin, usually with a lash at its apex (Fig. 1.38A). Multiple lesions may be present and occasionally abscesses may involve the entire lid margin. Treatment involves topical (occasionally oral) antibiotics, hot compresses and epilation of the associated lash.

Impetigo

Impetigo is an superficial skin infection caused by *Staphylococcus aureus* or *Streptococcus pyogenes*; it typically affects children. Involvement of the eyelids is usually associated with infection of the face. Painful erythematous macules rapidly develop into thin-walled blisters, which develop golden-yellow crusts on rupturing (Fig. 1.38B). There may be fever, malaise and local lymphadenopathy. Treatment is with topical and sometimes oral antibiotics (beta-lactamase resistant), and preventative measures to reduce transmission as the condition is highly contagious; it is particularly dangerous to neonates, contact with whom should be avoided.

Erysipelas

Erysipelas (St Anthony's fire) is an uncommon acute, potentially severe, dermal and superficial lymphatic infection usually caused by *S. pyogenes*. Diabetes, obesity and alcohol abuse are predisposing. An inflamed erythematous plaque develops (Fig. 1.38C); a well-defined raised border distinguishes erysipelas from other forms of cellulitis. Complications such as metastatic infection are rare. Treatment is with oral antibiotics, but recurrence is common.

Fig. 1.38 Bacterial infections. **(A)** External hordeolum (stye); **(B)** impetigo; **(C)** erysipelas; **(D)** necrotizing fasciitis

Necrotizing fasciitis

Necrotizing fasciitis is a rare but commonly very severe infection involving subcutaneous soft tissue and the skin, with associated rapidly progressive necrosis. It is usually caused by *S. pyogenes* and occasionally *S. aureus*. The most frequent sites of involvement are the extremities, trunk and perineum, as well as postoperative wound sites. Unless early aggressive treatment is instituted, in the form of surgical debridement and high-dose intravenous antibiotics, death may result. Periocular infection is rare; redness and oedema are followed by the formation of large bullae and black discoloration of the skin due to necrosis (Fig. 1.38D).

VIRAL INFECTIONS

Molluscum contagiosum

Introduction

Molluscum contagiosum is a skin infection caused by a human-specific double-stranded DNA poxvirus that typically affects otherwise healthy children, with a peak incidence between 2 and 4 years of age. Transmission is by contact and subsequently by autoinoculation. Multiple, and occasionally confluent, lesions may develop in immunocompromised patients. Histopathology shows a central pit and lobules of hyperplastic epidermis with intracytoplasmic (Henderson–Patterson) inclusion bodies that displace the nuclear remnant to the edge of the cell. The bodies are small and eosinophilic near the surface, and large and basophilic deeper down (Fig. 1.39A).

Diagnosis

Single or multiple pale, waxy, umbilicated nodules develop (Fig. 1.39B); white cheesy material consisting of infected degenerate cells can be expressed from the lesion. Lesions on the lid margin (Fig. 1.39C) may shed virus into the tear film and give rise to a secondary ipsilateral chronic follicular conjunctivitis. Unless the lid margin is examined carefully the causative molluscum lesion may be overlooked.

Treatment

Spontaneous resolution will usually occur within a few months so treatment may not be necessary, particularly in children, unless complications such as a significant secondary conjunctivitis are problematic. Options include shave excision, cauterization, chemical ablation, cryotherapy and pulsed dye laser.

Herpes zoster ophthalmicus

Herpes zoster ophthalmicus (HZO – Fig. 1.40) is a common, generally unilateral infection caused by varicella-zoster virus. It is discussed in detail in Ch. 6.

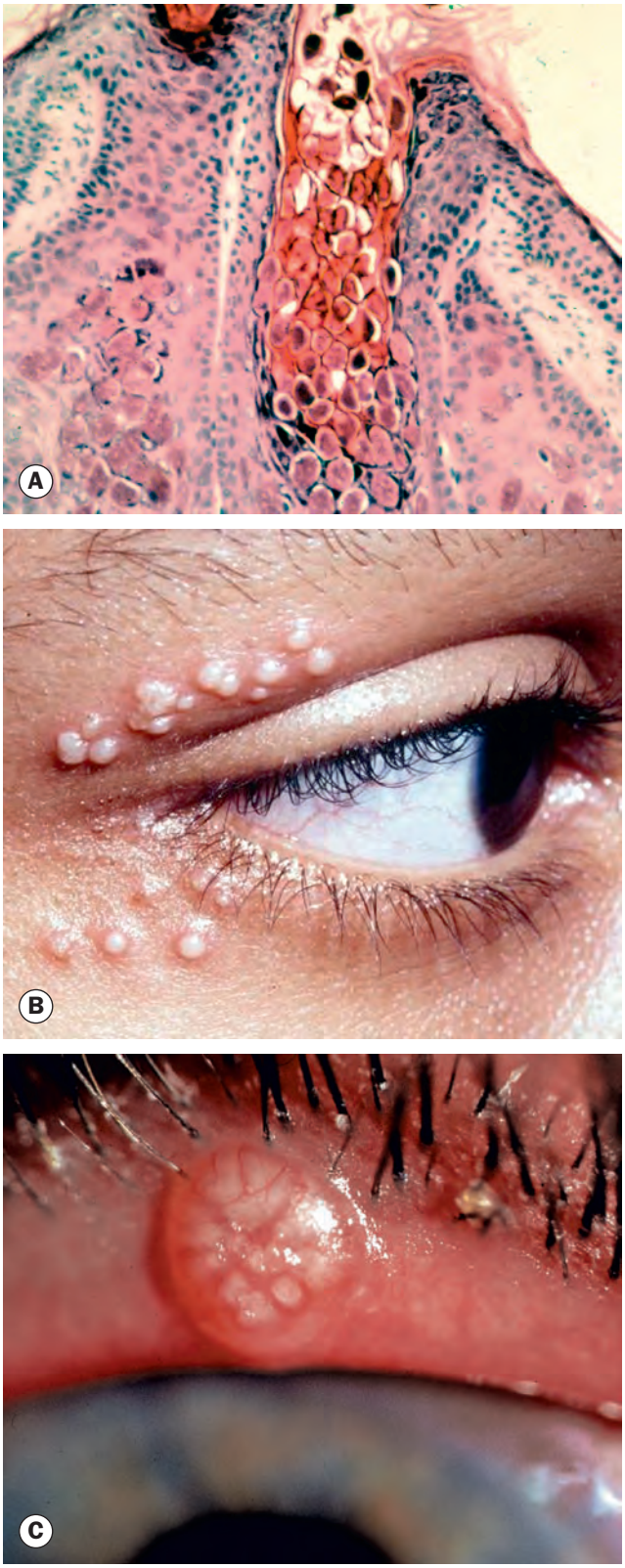


Fig. 1.39 Molluscum contagiosum. **(A)** Histopathology shows lobules of hyperplastic epidermis and a pit containing intracytoplasmic inclusion bodies; **(B)** multiple molluscum nodules; **(C)** lid margin nodule

(Courtesy of A Garner – fig. A; N Rogers – fig. B)

Fig. 1.40 Herpes zoster ophthalmicus – maculopapular crusting rash with periocular oedema, showing clear midline delineation

Herpes simplex

Introduction

Herpes simplex skin rash results from either primary infection or reactivation of herpes simplex virus previously dormant in the trigeminal ganglion. Prodromal facial and lid tingling lasting about 24 hours is followed by the development of eyelid and periocular skin vesicles (Fig. 1.41A) that break down over 48 hours (Fig. 1.41B). Although typically still confined to a single dermatome and with individual lesions that are often similar in appearance, the distribution of the herpes simplex skin rash contrasts with the sharply delineated unilateral involvement in HZO (see Fig. 1.40). There is commonly associated papillary conjunctivitis, discharge and lid swelling; dendritic corneal ulcers can develop, especially in atopic patients, in whom skin involvement can be extensive and very severe (eczema herpeticum – Fig. 1.41C).

Treatment

In many patients things will gradually settle without treatment over about a week. If treatment is necessary, a topical (aciclovir cream five times daily for 5 days) or oral (oral aciclovir, famciclovir or valaciclovir) antiviral agent can be used. Antibiotics (e.g. co-amoxiclav, erythromycin) may also be required in patients with secondary bacterial infection; this is particularly common in eczema herpeticum.

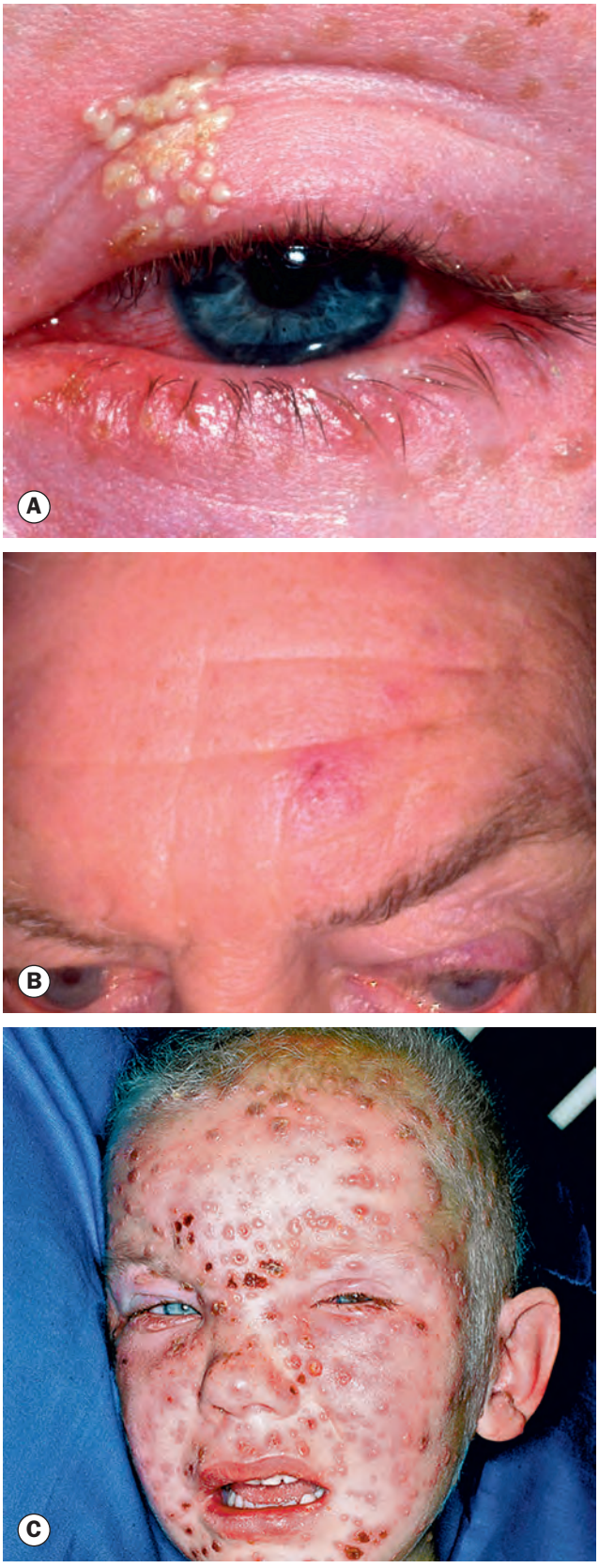


Fig. 1.41 Herpes simplex. **(A)** Vesicles; **(B)** progression to crusting; **(C)** eczema herpeticum

BLEPHARITIS

Chronic blepharitis

Introduction

Chronic blepharitis (chronic marginal blepharitis) is a very common cause of ocular discomfort and irritation. The poor correlation between symptoms and signs, the uncertain aetiology and mechanisms of the disease process all combine to make management difficult. Blepharitis may be subdivided into anterior and posterior, although there is considerable overlap and both types are often present (mixed blepharitis).

- Anterior blepharitis affects the area surrounding the bases of the eyelashes and may be staphylococcal or seborrhoeic. It is sometimes regarded as related more to chronic infective elements and hence more amenable to treatment and remission than the posterior form. An aetiological factor in staphylococcal blepharitis may be an abnormal cell-mediated response to components of the cell wall of *S. aureus,* which may also be responsible for the red eyes and peripheral corneal infiltrates seen in some patients; it is more common and more marked in patients with atopic dermatitis. Seborrhoeic blepharitis is strongly associated with generalized seborrhoeic dermatitis that characteristically involves the scalp, nasolabial folds, skin behind the ears and the sternum.
- Posterior blepharitis is caused by meibomian gland dysfunction and alterations in meibomian gland secretions. Bacterial lipases may result in the formation of free fatty acids. This increases the melting point of the meibum, preventing its expression from the glands, contributing to ocular surface irritation and possibly enabling growth of *S. aureus*. Loss of the tear film phospholipids that act as surfactants results in increased tear evaporation and osmolarity, and an unstable tear film. Posterior blepharitis is commonly thought of as a more persistent and chronic inflammatory condition than anterior blepharitis; there is an association with acne rosacea.
- A reaction to the extremely common hair follicle and sebaceous gland-dwelling mite *Demodex* and other microorganisms may play a causative role in some patients – *Demodex folliculorum longus* in anterior blepharitis and *Demodex folliculorum brevis* in posterior blepharitis – though the mite can be found normally in a majority of older patients, most of whom do not develop symptomatic blepharitis. It has been proposed that circumstances such as overpopulation or hypersensitivity (perhaps to a bacillus carried symbiotically by *Demodex*) may lead to symptoms. *Demodex* mites are a major cause of the animal disease mange.

The characteristics of the different forms of blepharitis are set out in Table 1.4.

Diagnosis

Involvement is usually bilateral and symmetrical.

- **Symptoms** are caused by disruption of normal ocular surface function and reduction in tear stability, and are similar in all forms of blepharitis, though stinging may be more common in posterior disease. Because of poor correlation between the severity of symptoms and signs it can be difficult to objectively assess the benefit of treatment. Burning, grittiness, mild photophobia, and crusting and redness of the lid margins with remissions and exacerbations are characteristic. Symptoms are usually worse in the mornings although in patients with associated dry eye they

Table 1.4 Summary of characteristics of chronic blepharitis

Feature		Anterior blepharitis		Posterior blepharitis
		Staphylococcal	Seborrhoeic	
Lashes	Deposit	Hard	Soft	
	Loss	++	+	
	Distorted or trichiasis	++	+	
Lid margin	Ulceration	+		
	Notching	+		++
Cyst	Hordeolum	++		
	Meibomian			++
Conjunctiva	Phlyctenule	+		
Tear film	Foaming			++
	Dry eye	+	+	++
Cornea	Punctate erosions	+	+	++
	Vascularization	+	+	++
	Infiltrates	+	+	++
Commonly associated skin disease		Atopic dermatitis	Seborrhoeic dermatitis	Acne rosacea

may increase during the day. Contact lens wear may be poorly tolerated.

- **Signs – staphylococcal blepharitis**
  - Hard scales and crusting mainly located around the bases of the lashes; collarettes are cylindrical collections around lash bases (Fig. 1.42A).
  - Mild papillary conjunctivitis and chronic conjunctival hyperaemia are common.
  - Long-standing cases may develop scarring and notching (tylosis) of the lid margin, madarosis, trichiasis and poliosis.
  - Associated tear film instability and dry eye syndrome are common.
  - Atopic keratoconjunctivitis may be present in patients with atopic dermatitis.
- **Signs – seborrhoeic blepharitis**
  - Hyperaemic and greasy anterior lid margins with soft scales and adherence of lashes to each other (Fig. 1.42B).

Fig. 1.42 Chronic anterior blepharitis. **(A)** Scales and crusting including collarettes in staphylococcal; **(B)** greasy lid margin with sticky lashes in seborrhoeic

- **Signs – posterior blepharitis** (meibomian gland disease)
  - Excessive and abnormal meibomian gland secretion, manifesting as capping of meibomian gland orifices with oil globules (Fig. 1.43A).
  - Pouting, recession, or plugging of meibomian gland orifices (Fig. 1.43B).
  - Hyperaemia and telangiectasis of the posterior lid margin.
  - Pressure on the lid margin results in expression of meibomian fluid that may be turbid or toothpaste-like (Fig. 1.43C); in severe cases the secretions become so inspissated that expression is impossible.
  - Lid transillumination may show gland loss and cystic dilatation of meibomian ducts.
  - The tear film is oily and foamy and often unstable, and froth may accumulate on the lid margins (Fig. 1.43D) or inner canthi.
- ***Demodex* infestation** may lead to cylindrical dandruff-like scaling (collarettes) around the base of eyelashes, though this is not always present. The mites can be demonstrated under ×16 slit lamp magnification by first manually clearing around the base of an eyelash then with fine forceps gently rotating the lash or moving it from side to side for 5–10 seconds, when if one or more mites (0.2–0.4 mm long – Fig. 1.44) does not emerge the lash should be gently epilated; slide microscopy can be performed on the mites or lashes if necessary.
- **Secondary changes** include papillary conjunctivitis, inferior corneal punctate epithelial erosions, corneal scarring and vascularization including Salzmann nodular degeneration and advancing wave-like epitheliopathy-type changes, stye formation, marginal keratitis, and occasionally bacterial keratitis (especially in contact lens wearers) and phlyctenulosis.

Treatment

There is limited evidence to support any particular treatment protocol for blepharitis. Patients should be advised that a permanent cure is unlikely, but control of symptoms is usually possible. The treatment of anterior and posterior disease is broadly similar for both types, particular given that they commonly co-exist, but some treatments are fairly specific for one or the other.

- **Lid hygiene** can be carried out once or twice daily initially; compliance and technique is highly variable.
  - A warm compress should first be applied for several minutes to soften crusts at the bases of the lashes.
  - Lid cleaning is subsequently performed to mechanically remove crusts and other debris, scrubbing the lid margins with a cotton bud or clean facecloth dipped in a warm dilute solution of baby shampoo or sodium bicarbonate.
  - Commercially produced soap/alcohol impregnated pads for lid scrubbing are available and are often highly effective, but care should be taken not to induce mechanical irritation.

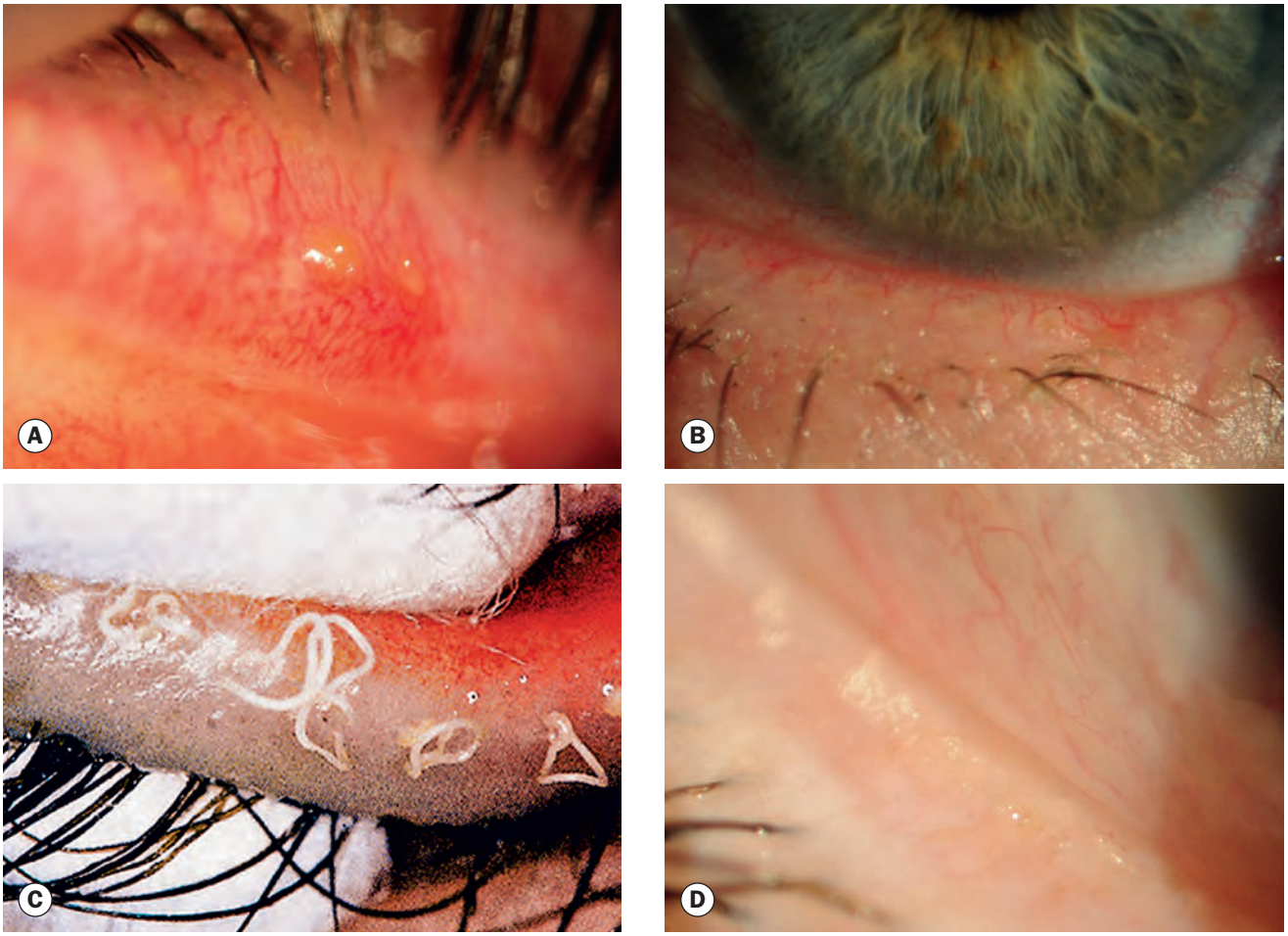


Fig. 1.43 Chronic posterior blepharitis. **(A)** Capping of meibomian gland orifices by oil globules; **(B)** hyperaemic, telangiectatic lid margin; **(C)** expressed toothpaste-like material; **(D)** froth on the eyelid margin

(Courtesy of J Silbert, from Anterior Segment Complications of Contact Lens Wear, *Butterworth-Heinemann 1999 – fig. C)*

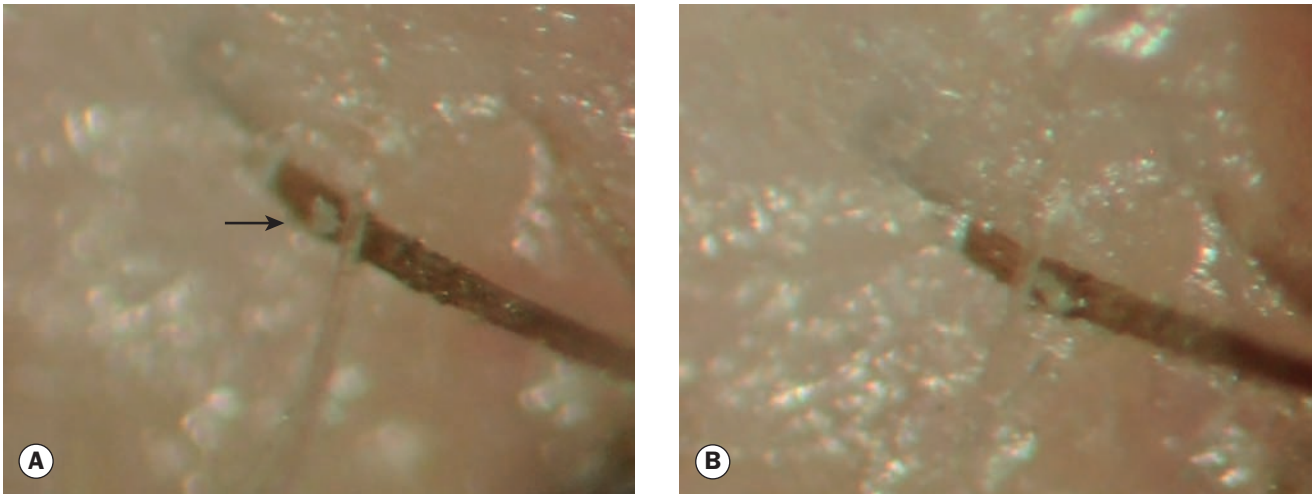


Fig. 1.44 *Demodex* mite. **(A)** Mite visible at eyelash base as a whitish lesion (arrow) after lash manipulation following clearance of collarette; **(B)** photograph taken two seconds later showing rapid migration

- When substantial meibomian gland disease is present, the regimen may include expression of accumulated meibum by rolling the finger anteriorly over the margin.
- The putative action of lid hygiene against *Demodex* is via prevention of reproduction.
- Lid hygiene can be performed less frequently as the condition is brought under control.

- **Antibiotics**
  - Topical sodium fusidic acid, erythromycin, bacitracin, azithromycin or chloramphenicol is used to treat active folliculitis in anterior disease and is occasionally used for an extended period. Following lid hygiene the ointment should be rubbed onto the anterior lid margin with a cotton bud or clean finger.
  - Oral antibiotic regimens include doxycycline (50–100 mg twice daily for 1 week and then daily for 6–24 weeks), other tetracyclines, or azithromycin (500 mg daily for 3 days for three cycles at 1-week intervals); antibiotics are thought to reduce bacterial colonization and may also exert other effects such as a reduction in staphylococcal lipase production with tetracyclines. Tetracyclines may be more effective in the treatment of posterior disease, and azithromycin in anterior. Tetracyclines should not be used in children under the age of 12 years or in pregnant or breast-feeding women because they are deposited in growing bone and teeth; patients should also be aware of the possibility of increased sun sensitivity. Erythromycin 250 mg once or twice daily is an alternative.
- **Plant and fish oil supplements** have been shown to be of substantial benefit in some cases.
- **Topical steroid.** A low potency preparation such as fluorometholone 0.1% or loteprednol four times daily for 1 week is useful in patients with substantial active inflammation, especially papillary conjunctivitis; occasionally a higher strength preparation is used.
- **Tear substitutes** and other dry eye treatments are typically helpful for associated tear insufficiency and instability.
- **Tea tree oil** has been suggested as a treatment, based primarily on its likely activity against *Demodex* infestation; the optimal vehicle and regimen has not been established but lid, eyebrow and periocular skin cleansing once daily with a 50% scrub and application of 5% ointment has been described. Topical permethrin and topical (1% cream) or oral (two doses of 200 µg/kg 1 week apart) ivermectin have also been used by some practitioners. High temperature cleaning of bedding, the use of tea tree shampoo and facial soap, and treating the patient's partner may all help to reduce recurrences.
- **Novel therapies** include topical ciclosporin, pulsed light application, and purpose-designed devices to probe, heat and/or express the meibomian glands (e.g. Lipiflow ™) in posterior disease.
- **Complications** are treated specifically.

Phthiriasis palpebrarum

The crab louse *Phthirus pubis* is adapted to living in pubic hair, but is also commonly found in other hair-covered body areas such as the chest, axillae and eyelids (phthiriasis palpebrarum). Symptoms consist of chronic irritation and itching of the lids, but the lice are often an incidental discovery. Conjunctivitis is uncommon. The lice are readily visible anchored to lashes (Fig. 1.45A); lice have six legs rather than the eight possessed by ticks (see next). Ova and their empty shells appear as oval, brownish, opalescent pearls adherent to the base of the cilia (Fig. 1.45B). Treatment consists of mechanical removal of the lice and their attached lashes with fine forceps. If necessary, topical yellow mercuric oxide 1% or petroleum jelly can be applied to the lashes and lids twice a day for 10 days. Delousing of the patient, family members, clothing and bedding is important to prevent recurrence.

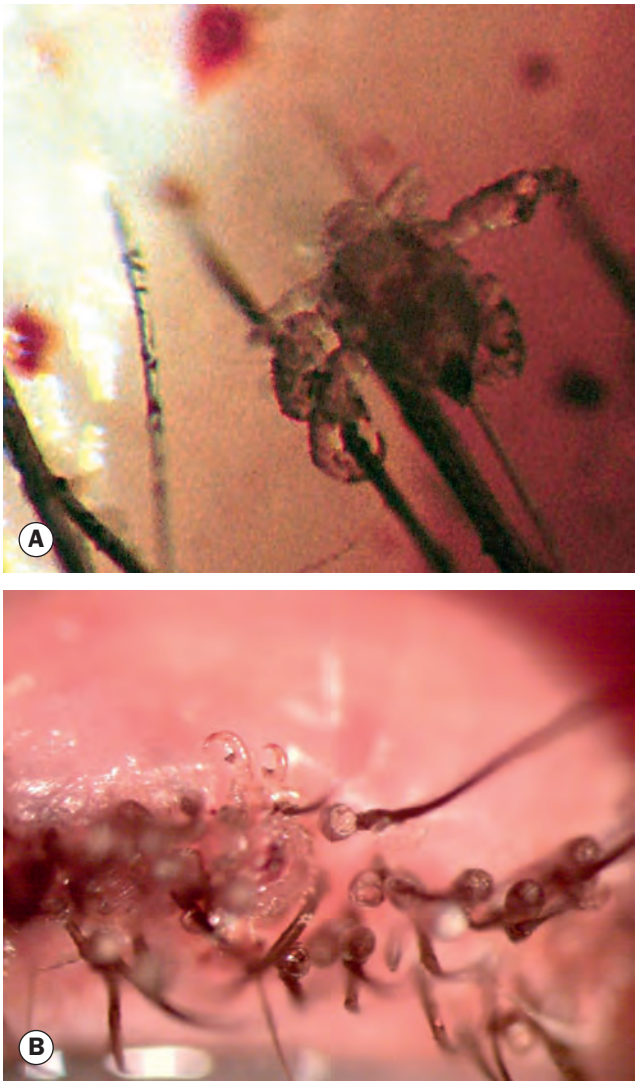


Fig. 1.45 Phthiriasis palpebrarum. **(A)** Louse anchored to lashes; **(B)** louse, ova and shells
(Courtesy of D Smit – fig. B)

Tick infestation of the eyelid

Ticks (see Fig. 11.69B) can attach themselves to the eyelid and should be removed at the earliest opportunity in order to minimize the risk of contracting a tick-borne zoonosis such as Lyme disease, Rocky Mountain fever or tularaemia. If the tick is attached some distance from the eye such that spray can safely be applied, an insect repellent containing pyrethrin or a pyrethroid should be sprayed on the tick twice at intervals of a minute; alternatively a scabies cream containing permethrin can be applied. These have a toxic effect that prevents the tick from injecting saliva, and after 24 hours it should drop off or can be removed with fine-tipped forceps at the slit lamp (blunt-tipped needle-holders are an alternative in restrained small children). It is critical that the tick is detached as close to its skin attachment as possible in order to remove its head and mouthparts, following which it might be retained in sealed packaging to permit identification if necessary. In areas endemic for Lyme disease, some authorities suggest routine antibiotic prophylaxis with doxycycline (in the absence of contraindications) following a confirmed deer tick bite, but as a minimum patients should be told to seek medical advice urgently at the onset of suspicious symptoms, particularly erythema migrans, over the subsequent few weeks. Lyme disease transmission is thought to require attachment of the tick for at least 36 hours.

Angular blepharitis

The infection is usually caused by *Moraxella lacunata* or *S. aureus* although other bacteria, and rarely herpes simplex, have also been implicated. Red, scaly, macerated and fissured skin is seen at the lateral and/or medial canthi of one or both eyes (Fig. 1.46A). Skin chafing secondary to tear overflow, especially at the lateral canthus, can cause a similar clinical picture, and may also predispose to infection (Fig. 1.46B). Associated papillary and follicular conjunctivitis may occur. Treatment involves topical chloramphenicol, bacitracin or erythromycin.

Childhood blepharokeratoconjunctivitis

Childhood blepharokeratoconjunctivitis is a poorly defined condition that tends to be more severe in Asian and Middle Eastern populations. Presentation is usually at about 6 years of age with recurrent episodes of anterior or posterior blepharitis, sometimes associated with recurrent styes or chalazia. Constant eye rubbing and photophobia may lead to misdiagnosis as allergic eye disease. Conjunctival changes include diffuse hyperaemia, bulbar phlyctens and follicular or papillary hyperplasia.

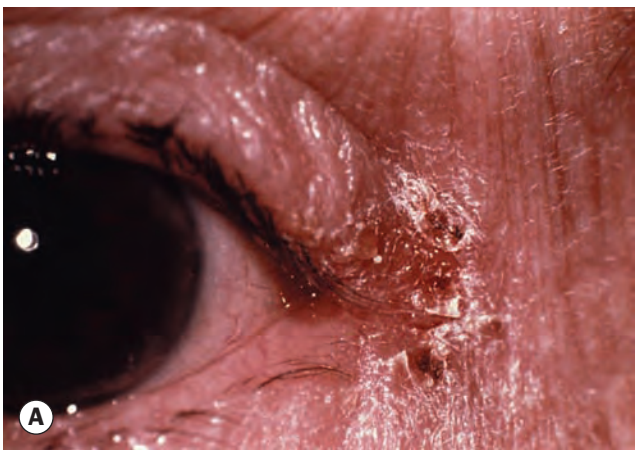
Corneal changes include superficial punctate keratopathy, marginal keratitis, peripheral vascularization and axial subepithelial haze. Treatment is with lid hygiene and topical antibiotic ointment at bedtime. Topical low-dose steroids (prednisolone 0.1% or fluorometholone 0.1%) and erythromycin syrup 125 mg daily for 4–6 weeks may also be used.

PTOSIS

Classification

Ptosis is an abnormally low position of the upper lid; it may be congenital or acquired.

- **Neurogenic** ptosis is caused by an innervational defect such as third nerve paresis and Horner syndrome (see Ch. 19).
- **Myogenic** ptosis is caused by a myopathy of the levator muscle itself, or by impairment of transmission of impulses at the neuromuscular junction (neuromyopathic). Acquired myogenic ptosis occurs in myasthenia gravis, myotonic dystrophy and progressive external ophthalmoplegia (see Ch. 19).
- **Aponeurotic** or involutional ptosis is caused by a defect in the levator aponeurosis.
- **Mechanical** ptosis is caused by the gravitational effect of a mass or by scarring.



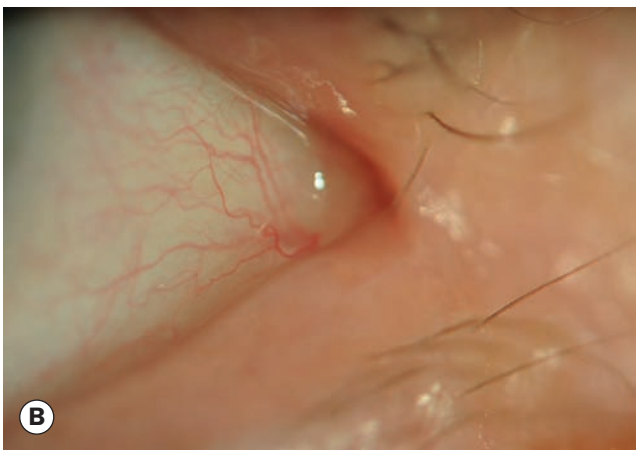


Fig. 1.46 **(A)** Angular blepharitis; **(B)** tear overflow from a lax and rounded lateral canthus

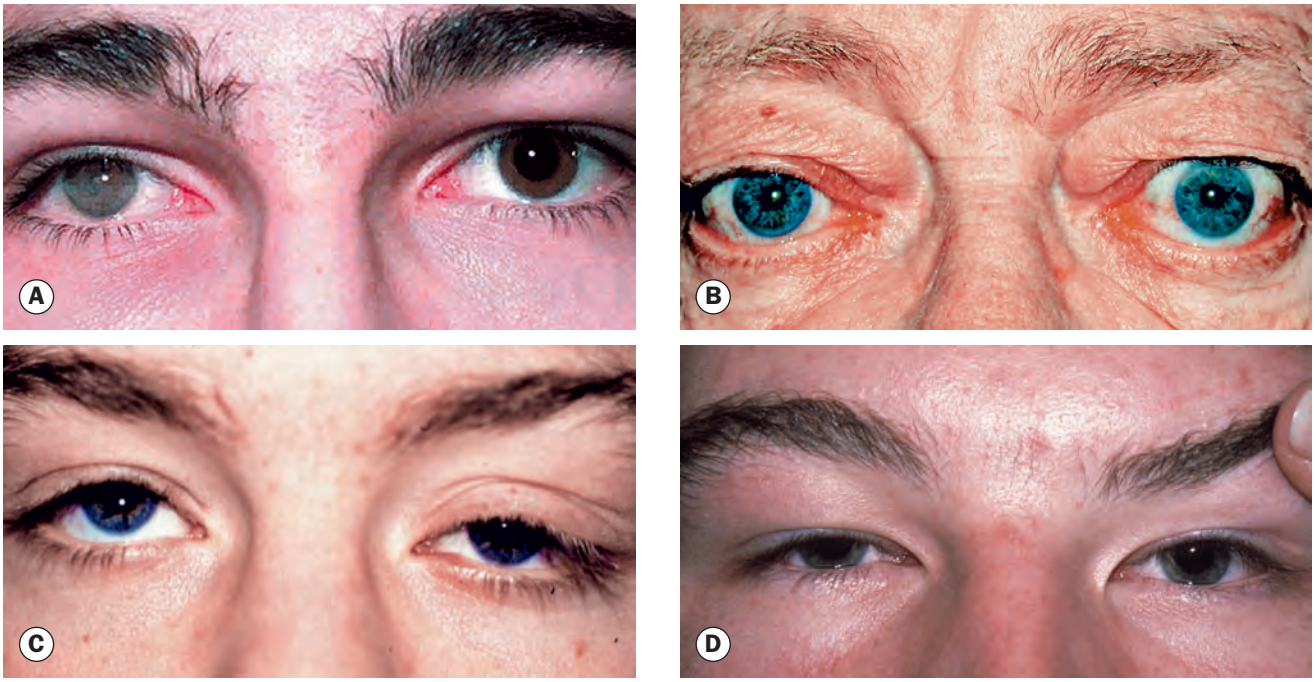


Fig. 1.47 Causes of pseudoptosis. **(A)** Right phthisis bulbi; **(B)** contralateral lid retraction; **(C)** ipsilateral hypotropia; **(D)** bilateral brow ptosis
(Courtesy of S Webber – figs C and D)

Clinical evaluation

General

The age at onset of ptosis and its duration will usually distinguish congenital from acquired cases. If the history is ambiguous, old photographs may be helpful. It is also important to enquire about symptoms of possible underlying systemic disease, such as associated diplopia, variability of ptosis during the day and excessive fatigue.

Pseudoptosis

A false impression of ptosis may be caused by the following:

- **Lack of support** of the lids by the globe may be due to an orbital volume deficit associated with an artificial eye, microphthalmos, phthisis bulbi (Fig. 1.47A), or enophthalmos.
- **Contralateral lid retraction**, which is detected by comparing the levels of the upper lids, remembering that the margin of the upper lid normally covers the superior 2 mm of the cornea (Fig. 1.47B).
- **Ipsilateral hypotropia** causes pseudoptosis because the upper lid follows the globe downwards (Fig. 1.47C). It disappears when the hypotropic eye assumes fixation on covering the normal eye.
- **Brow ptosis** due to excessive skin on the brow, or seventh nerve palsy, which is diagnosed by manually elevating the eyebrow (Fig. 1.47D).
- **Dermatochalasis.** Overhanging skin on the upper lids (Fig. 1.48) may be mistaken for ptosis, but may also cause mechanical ptosis.

Measurements

- **Margin–reflex distance** is the distance between the upper lid margin and the corneal reflection of a pen torch held by the

Fig. 1.48 Marked right dermatochalasis and brow ptosis
(Courtesy of S Chen)

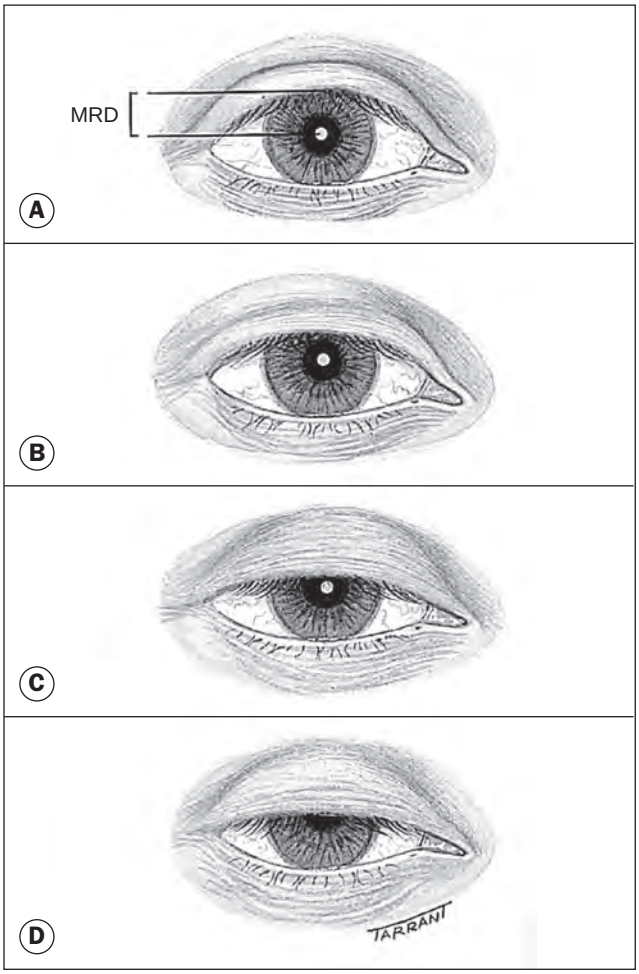


Fig. 1.49 Margin–reflex distance. **(A)** Normal; **(B)** mild ptosis; **(C)** moderate ptosis; **(D)** severe ptosis

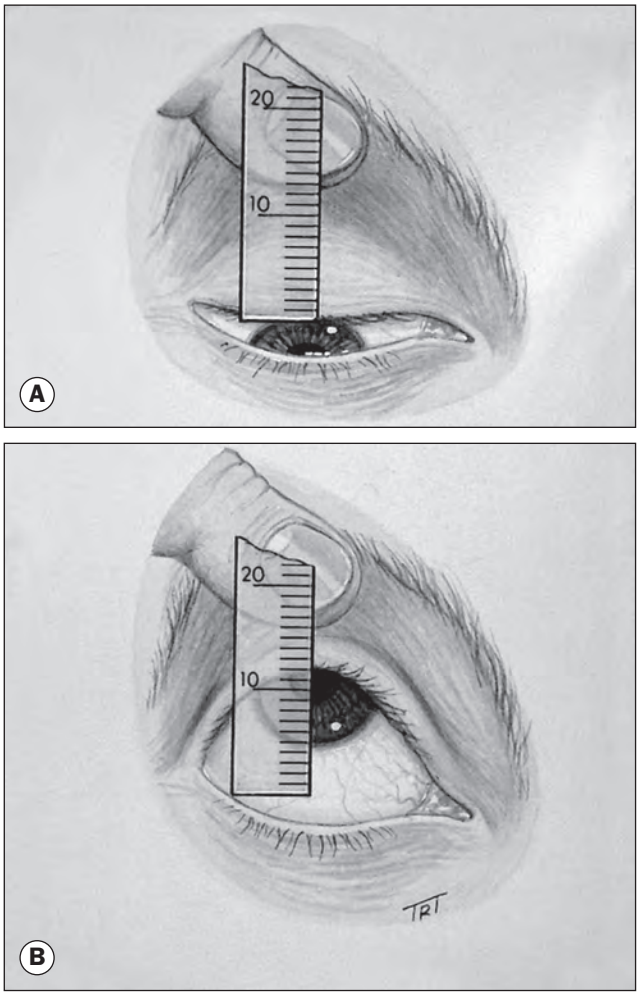


Fig. 1.51 Measurement of levator function

examiner on which the patient fixates (Fig. 1.49); the normal measurement is 4–5 mm.

- **Palpebral fissure height** is the distance between the upper and lower lid margins, measured in the pupillary plane (Fig. 1.50). The upper lid margin normally rests about 2 mm below the upper limbus and the lower 1 mm above the lower limbus. This measurement is shorter in males (7–10 mm) than in females (8–12 mm). Unilateral ptosis can be quantified by comparison with the contralateral side. Ptosis may be graded as mild (up to 2 mm), moderate (3 mm) and severe (4 mm or more).
- **Levator function** (upper lid excursion) is measured by placing a thumb firmly against the patient's brow to negate the action of the frontalis muscle, with the eyes in downgaze (Fig. 1.51A). The patient then looks up as far as possible and the amount of excursion is measured with a rule (Fig. 1.51B). Levator function is graded as normal (15 mm or more), good (12–14 mm), fair (5–11 mm) and poor (4 mm or less).
- **Upper lid crease** is taken as the vertical distance between the lid margin and the lid crease in downgaze. In females it measures about 10 mm and in males 8 mm. Absence of the crease in a patient with congenital ptosis is evidence of poor levator function, whereas a high crease suggests an aponeurotic defect (usually involutional). The skin crease is also used as a guide to the initial incision in some surgical procedures.
- **Pretarsal show** is the distance between the lid margin and the skin fold with the eyes in the primary position.

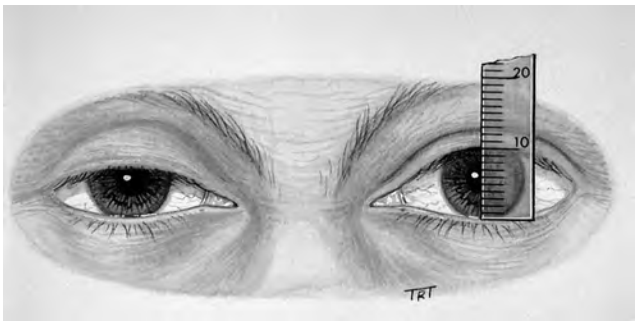


Fig. 1.50 Measurement of palpebral fissure height

Associated signs

- **The pupils** should be examined to exclude Horner syndrome and a subtle pupil-involving third nerve palsy – the latter is an unlikely acute clinical presentation (see Ch. 19).
- **Increased innervation** may flow to the levator muscle of a unilateral ptosis, particularly in upgaze. Associated increased innervation to the contralateral normal levator will result in lid retraction. The examiner should therefore manually elevate the ptotic lid and look for drooping of the opposite lid. If this occurs, the patient should be warned that surgical correction may induce a lower position in the opposite lid.
- **Fatigability** is tested by asking the patient to look up without blinking for 30–60 seconds. Progressive drooping of one or both lids, or an inability to maintain upgaze, is suggestive of myasthenia gravis (see Ch. 19). Myasthenic ptosis may show an overshoot of the upper lid on saccade from downgaze to the primary position (Cogan twitch sign) and a 'hop' on side-gaze.
- **Ocular motility defects**, particularly of the superior rectus, must be evaluated in patients with congenital ptosis. Correction of an ipsilateral hypotropia may improve the degree of ptosis. Deficits consistent with a subtle or partial third nerve paresis should be identified.
- **Jaw-winking** can be identified by asking the patient to chew and move the jaws from side to side (see below).
- **The Bell phenomenon** is tested by manually holding the lids open, asking the patient to try to shut the eyes and observing upward and outward rotation of the globe. A weak Bell phenomenon carries a variable risk of postoperative exposure keratopathy, particularly following large levator resections or suspension procedures.
- **The tear film** should be inspected – a poor volume or unstable film may be worsened by ptosis surgery and should be addressed preoperatively as far as possible.

Simple congenital ptosis

Diagnosis

Congenital ptosis probably results from a failure of neuronal migration or development with muscular sequelae secondary to this; a minority of patients have a family history.

- **Signs** (Fig. 1.52)
  - Unilateral or bilateral ptosis of variable severity.
  - Absent upper lid crease and poor levator function.
  - In downgaze the ptotic lid is higher than the normal because of poor relaxation of the levator muscle. This is in contrast to acquired ptosis, in which the affected lid is either level with or lower than the normal lid on downgaze.
  - Following surgical correction the lid lag in downgaze may worsen.
- **Associations**
  - Superior rectus weakness may be present because of its close embryological association with the levator.
  - Compensatory chin elevation in severe bilateral cases.
  - Refractive errors are common and more frequently responsible for amblyopia than the ptosis itself.

Treatment

Treatment should be carried out during the preschool years once accurate measurements can be obtained, but may be considered earlier in severe cases to prevent amblyopia. Levator resection (see below) is usually required.

Marcus Gunn jaw-winking syndrome

Introduction

About 5% of all cases of congenital ptosis are associated with the Marcus Gunn jaw-winking phenomenon. The vast majority are unilateral. Although the exact aetiology is unclear, it has been postulated that a branch of the mandibular division of the fifth cranial nerve is misdirected to the levator muscle.

Diagnosis

- **Signs**
  - Retraction of the ptotic lid in conjunction with stimulation of the ipsilateral pterygoid muscles by chewing, sucking, opening the mouth (Figs 1.53A and B) or contralateral jaw movement.
  - Less common stimuli to winking include jaw protrusion, smiling, swallowing and clenching of teeth.
  - Jaw-winking does not improve with age (Figs 1.53C and D), although patients may learn to mask it.

Treatment

Surgery should be considered if jaw-winking or ptosis represents a significant functional or cosmetic problem.

- **Mild cases** with reasonable levator function of 5 mm or better, and little synkinetic movement may be treated with unilateral levator advancement.
- **Moderate cases.** Unilateral levator disinsertion can be performed to address the synkinetic winking component, with ipsilateral brow (frontalis) suspension so that lid elevation is due solely to frontalis muscle elevation.
- **Bilateral surgery.** Bilateral levator disinsertion with bilateral brow suspension may be carried out to produce a symmetrical result.

Third nerve misdirection syndromes

Third nerve misdirection syndromes may be congenital, but more frequently follow acquired third nerve palsy. Bizarre movements of the upper lid accompany various eye movements (Fig. 1.54). Ptosis may also occur following aberrant facial nerve regeneration. Treatment is by levator disinsertion and brow suspension.

Fig. 1.52 Congenital ptosis. **(A)** Mild right ptosis; **(B)** good levator function; **(C)** severe left ptosis with absent skin crease; **(D)** very poor levator function; **(E)** severe bilateral ptosis; **(F)** very poor levator function

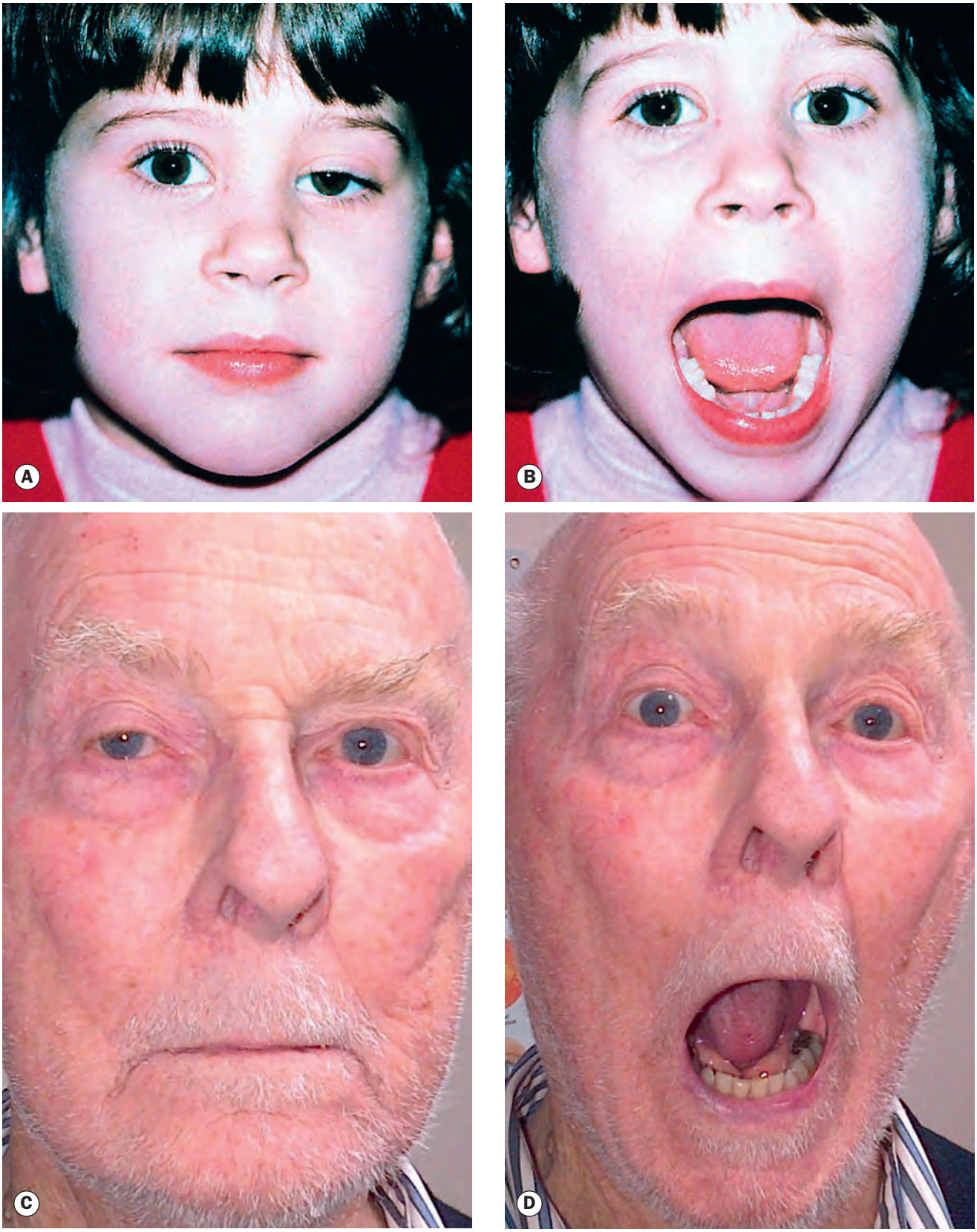


Fig. 1.53 Marcus Gunn jaw-winking syndrome. **(A)** Moderate left ptosis; **(B)** retraction of the lid on opening the mouth; **(C)** and **(D)** similar phenomenon in an older patient

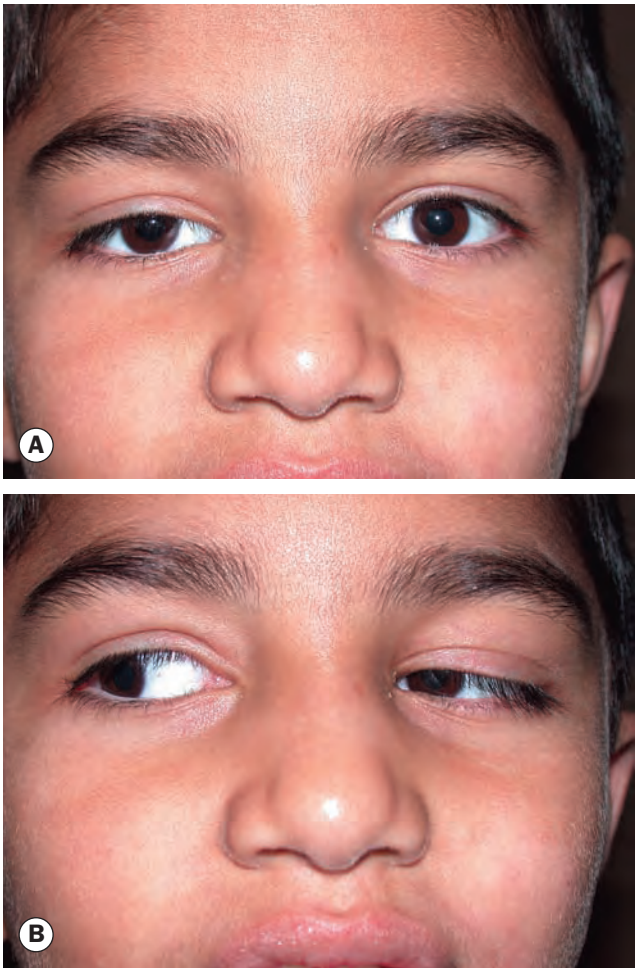


Fig. 1.54 Third nerve redirection. **(A)** Moderate right ptosis; **(B)** retraction of the lid on right gaze
(Courtesy of A Pearson)

Involutional ptosis

Involutional (aponeurotic) ptosis is an age-related condition caused by dehiscence, disinsertion or stretching of the levator aponeurosis, limiting the transmission of force from a normal levator muscle to the upper lid. Due to fatigue of the Müller muscle it frequently worsens towards the end of the day, so that it can sometimes be confused with myasthenic ptosis. There is a variable, usually bilateral, ptosis with a high upper lid crease and good levator function. In severe cases the upper lid crease may be absent, the eyelid above the tarsal plate very thin and the upper sulcus deep (Fig. 1.55). Treatment options include levator resection, advancement with reinsertion or anterior levator repair.

Mechanical ptosis

Mechanical ptosis is the result of impaired mobility of the upper lid. It may be caused by dermatochalasis, large tumours such as neurofibromas (Fig. 1.56), heavy scar tissue, severe oedema and anterior orbital lesions.

Fig. 1.55 Severe bilateral involutional ptosis with absent skin creases and deep sulci

Surgery

Anatomy

- **The levator aponeurosis** fuses with the orbital septum about 4 mm above the superior border of the tarsal plate (Fig. 1.57). Its posterior fibres insert into the lower third of the anterior surface of the tarsal plate. The medial and lateral horns are expansions that act as check ligaments. Surgically, the aponeurosis can be approached through the skin or conjunctiva.
- **Müller muscle** is inserted into the upper border of the tarsal plate and can be approached transconjunctivally.
- **The inferior tarsal aponeurosis** consists of the capsulopalpebral expansion of the inferior rectus muscle and is analogous to the levator aponeurosis.
- **The inferior tarsal muscle** is analogous to Müller muscle.

Conjunctiva–Müller resection

This involves excision of Müller muscle and overlying conjunctiva (Fig. 1.58A) with reattachment of the resected edges (Fig. 1.58B).

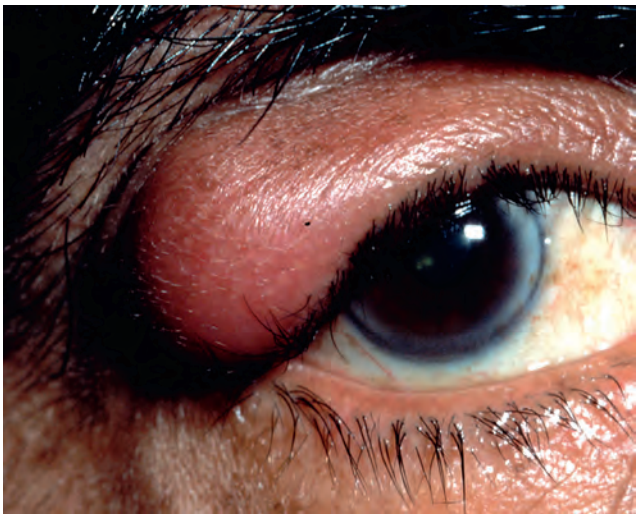


Fig. 1.56 Mechanical ptosis due to a neurofibroma

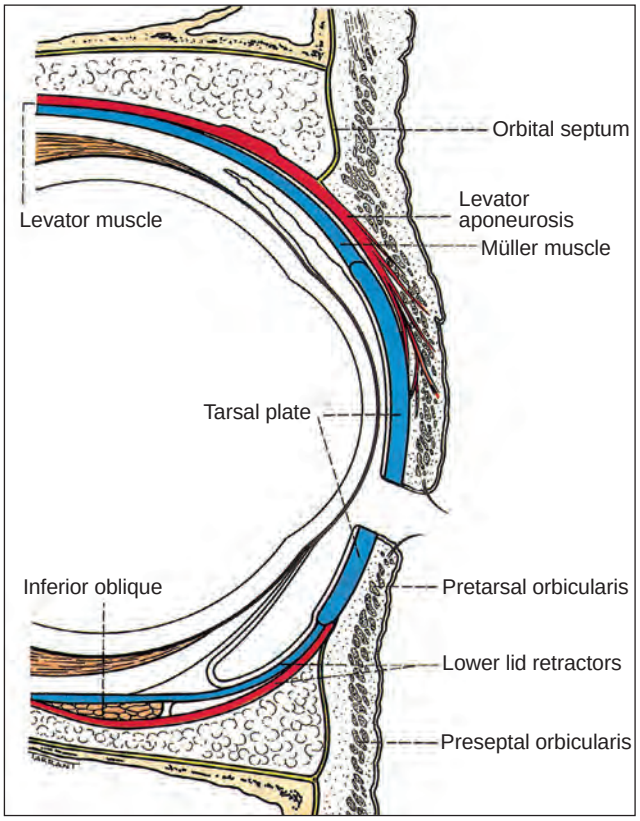


Fig. 1.57 Anatomy of the eyelid

The maximal elevation achievable is 2–3 mm, so it is used in cases of mild ptosis with good (at least 10 mm) levator function, which includes most cases of Horner syndrome and mild congenital ptosis.

Levator advancement (resection)

In this technique the levator complex is shortened through either an anterior – skin (Fig. 1.59) – or posterior – conjunctival – approach. Indications include ptosis of any cause, provided residual levator function is at least 5 mm. The extent of resection is determined by the severity of the ptosis and the amount of levator function.

Brow (frontalis) suspension

Brow (frontalis) suspension is used for severe ptosis (>4 mm) with very poor levator function (<4 mm) from a variety of causes, typical indications being ptosis associated with third nerve palsy, blepharophimosis syndrome and following an unsatisfactory result from previous levator resection. The tarsal plate is suspended from the frontalis muscle with a sling consisting of autologous fascia lata (Fig. 1.60) or non-absorbable material such as prolene or silicone.

ECTROPION

Involutional ectropion

Introduction

Involutional (age-related) ectropion affects the lower lid of elderly patients. It causes epiphora (tear overflow) and may exacerbate ocular surface disease. The red appearance of the exposed conjunctiva is cosmetically poor. In long-standing cases the tarsal conjunctiva may become chronically inflamed, thickened and keratinized (Fig. 1.61). Aetiological factors include:

- **Horizontal lid laxity** can be demonstrated by pulling the central part of the lid 8 mm or more from the globe, with a failure to snap back to its normal position on release without the patient first blinking.

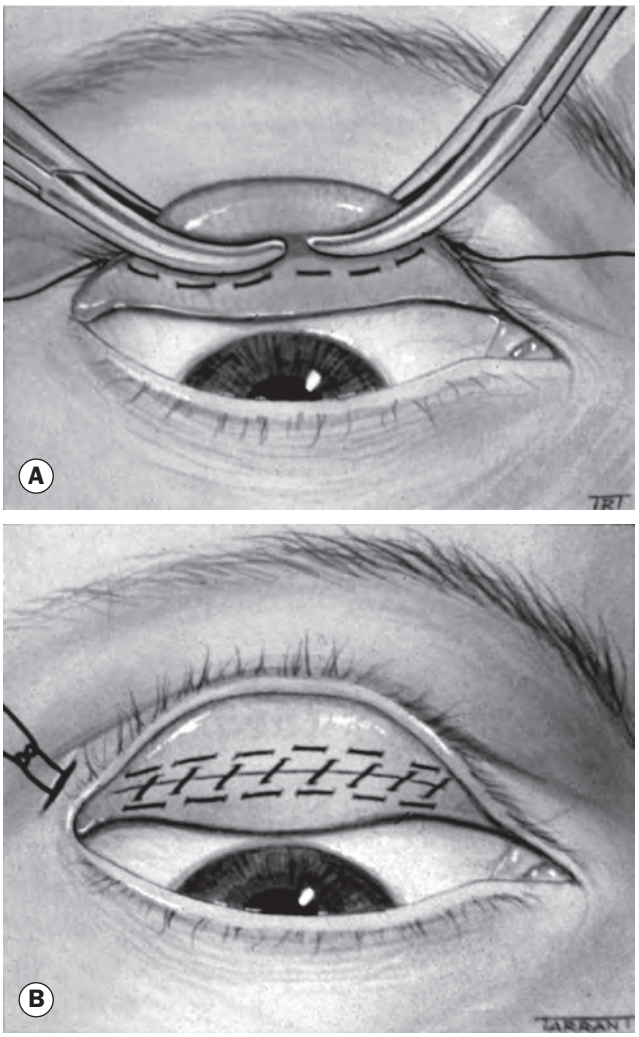
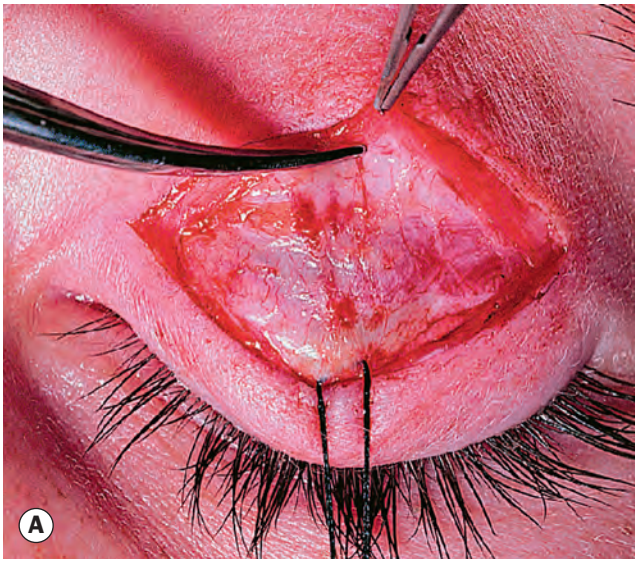
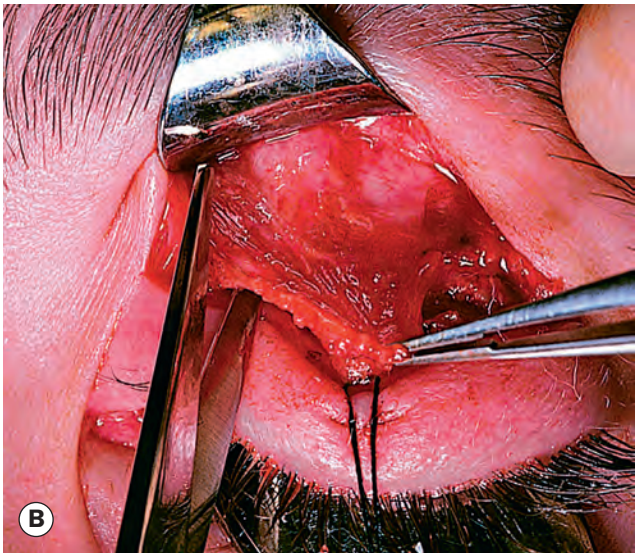


Fig. 1.58 Conjunctiva–Müller resection. **(A)** Clamping of conjunctiva and Müller muscle; **(B)** appearance after excision and suturing





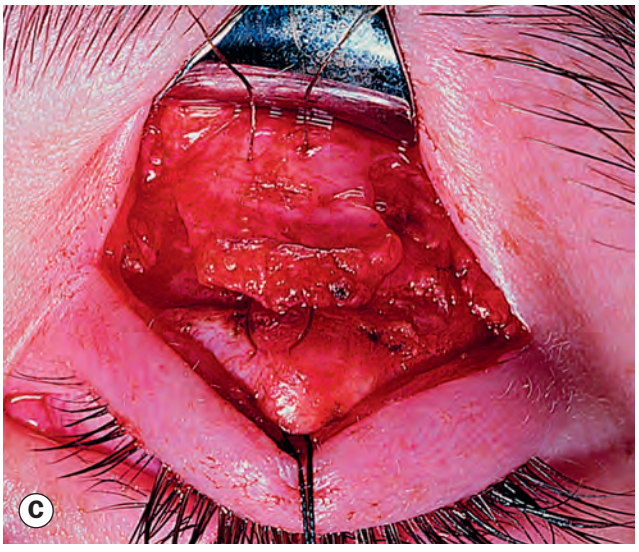
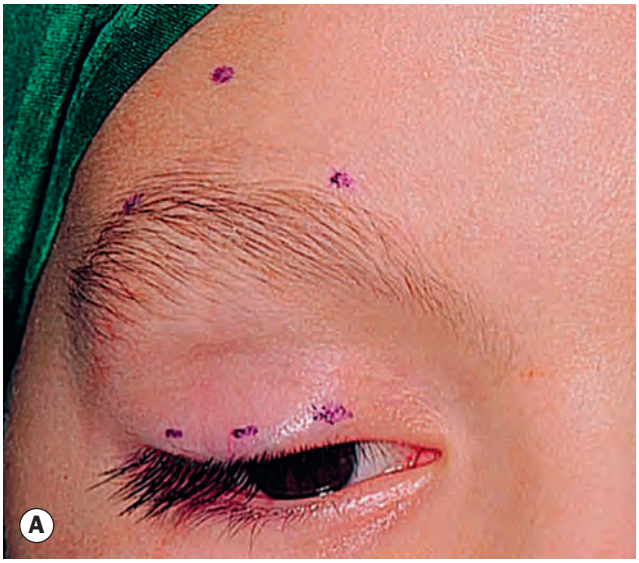


Fig. 1.59 Anterior levator resection. **(A)** Skin incision; **(B)** dissection and resection of levator aponeurosis; **(C)** levator reattachment to the tarsal plate

(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001)*



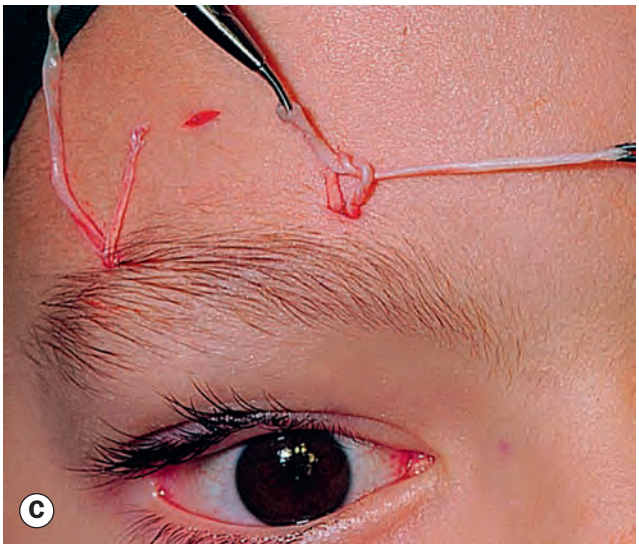


Fig. 1.60 Brow suspension. **(A)** Site of incisions marked; **(B)** threading of fascia lata strips; **(C)** tightening and tying of strips

(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001)*

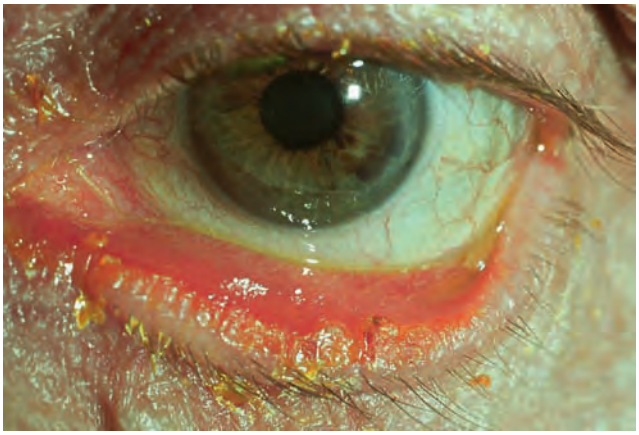


Fig. 1.61 Severe long-standing involutional ectropion with keratinization of the marginal conjunctiva
(Courtesy of C Barry)

- **Lateral canthal tendon laxity**, characterized by a rounded appearance of the lateral canthus (see Fig. 1.46B) and the ability to pull the lower lid medially more than 2 mm.
- **Medial canthal tendon laxity**, demonstrated by pulling the lower lid laterally and observing the position of the inferior punctum. If the lid is normal the punctum should not be displaced more than 1–2 mm. If laxity is mild the punctum reaches the limbus, and if severe it may reach the pupil.

Treatment

The approach to repair depends on apparent causation and the predominant location of the ectropion.

- **Generalized** ectropion is treated with repair of horizontal lid laxity. This is achieved with a lateral tarsal strip procedure, in which the lower canthal tendon is tightened by shortening and reattachment to the lateral orbital rim (Fig. 1.62); this is particularly helpful if the lateral canthus is rounded and lax, with associated tear overflow. Excision of a tarsoconjunctival pentagon (Fig. 1.63) is an alternative that can be placed to excise an area of misdirected lashes or keratinized conjunctiva.
- **Medial** ectropion, if mild, may be treated with a medial conjunctival diamond excision (medial spindle procedure), though must often be combined with a tarsal strip or lateral canthal sling (see 'Treatment'), or pentagon excision as significant horizontal laxity frequently co-exists.
- **Medial canthal tendon laxity**, if marked, requires stabilization prior to horizontal shortening to avoid excessive dragging of the punctum laterally.
- **Punctal ectropion** without more extensive lid involvement is considered in Ch. 2.

Cicatricial ectropion

Cicatricial ectropion is caused by scarring or contracture of the skin and underlying tissues, which pulls the eyelid away from the globe (Fig. 1.64). If the skin is pushed up over the orbital margin with a finger the ectropion will be relieved. Opening the mouth tends to accentuate the eversion. Depending on the cause, both lids may be involved and the defect may be local (e.g. trauma) or general (e.g. burns, dermatitis, ichthyosis). Mild localized cases are treated by excision of the offending scar tissue combined with a procedure that lengthens vertical skin deficiency, such as Z-plasty. Severe generalized cases require transposition flaps or free skin grafts; sources of skin include the upper lids, posterior auricular, preauricular and supraclavicular areas.

Paralytic ectropion/facial nerve palsy

Introduction

Paralytic ectropion is caused by ipsilateral facial nerve palsy (Fig. 1.65) and is associated with retraction of the upper and lower lids and brow ptosis; the latter may mimic narrowing of the palpebral aperture.

Complications include exposure keratopathy due to lagophthalmos, and watering caused by malposition of the inferior lacrimal punctum, failure of the lacrimal pump mechanism and an increase in tear production resulting from corneal exposure.

Treatment

- **Temporary** measures may be instituted to protect the cornea in anticipation of spontaneous recovery of facial nerve function.
  - Lubrication with higher viscosity tear substitutes during the day, with instillation of ointment and taping shut of the lids during sleep, are usually adequate in mild cases.
  - Botulinum toxin injection into the levator to induce temporary ptosis.
  - Temporary tarsorrhaphy may be necessary, particularly in patients with a poor Bell phenomenon with the cornea remaining exposed when the patient attempts to blink; the lateral aspects of the upper and lower lids are sutured together.
- **Permanent** treatment should be considered when there is irreversible damage to the facial nerve as may occur following removal of an acoustic neuroma, or when no further improvement has occurred for 6-12 months in a Bell palsy.
  - Medial canthoplasty may be performed if the medial canthal tendon is intact. The eyelids are sutured together medial to the lacrimal puncta (Fig. 1.66A) so that the puncta become inverted and the fissure between the inner canthus and puncta is shortened.
  - A lateral canthal sling or tarsal strip may be used to correct residual ectropion and raise the lateral canthus (Fig. 1.66B).
  - Upper eyelid lowering by levator disinsertion.
  - Gold weight implantation in the upper lid can assist closure.
  - A small lateral tarsorrhaphy is usually cosmetically acceptable.

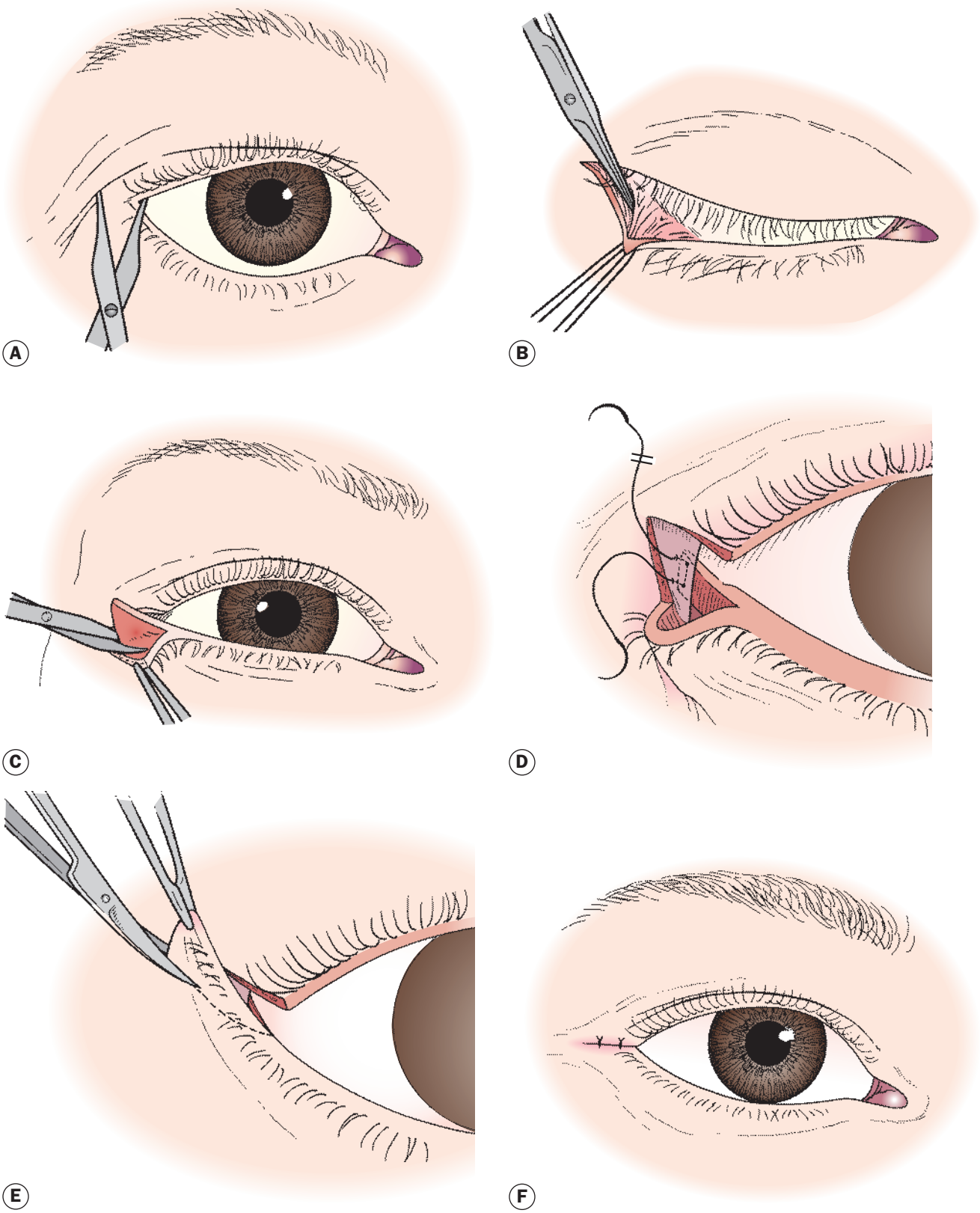


Fig. 1.62 Lateral tarsal strip procedure. **(A)** Lateral canthotomy; **(B)** cantholysis – the lower limb of the lateral canthal tendon is cut away from the inferior orbital rim; **(C)** the anterior and posterior lid lamellae are divided, and a 'strip' of tendon/lateral tarsal plate dissected out; **(D)** the strip is shortened and then reattached to the inner aspect of the orbital rim periosteum with 4-0 absorbable suture; **(E)** excess lid margin is trimmed; **(F)** the skin incision (canthotomy) is closed
(Courtesy of JA Nerad, from Techniques in Ophthalmic Plastic Surgery, *Saunders 2010)*

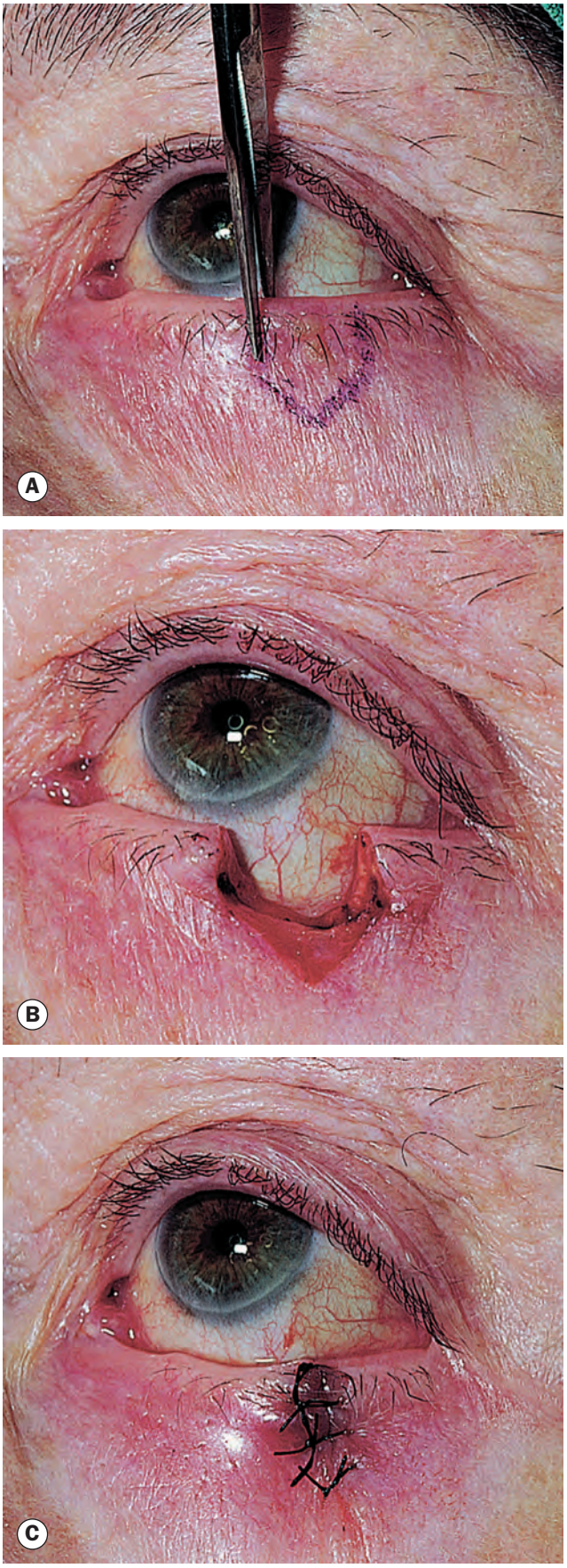


Fig. 1.63 Horizontal lid shortening to correct ectropion. **(A)** Marking; **(B)** excision of a pentagon; **(C)** closure

(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001)*

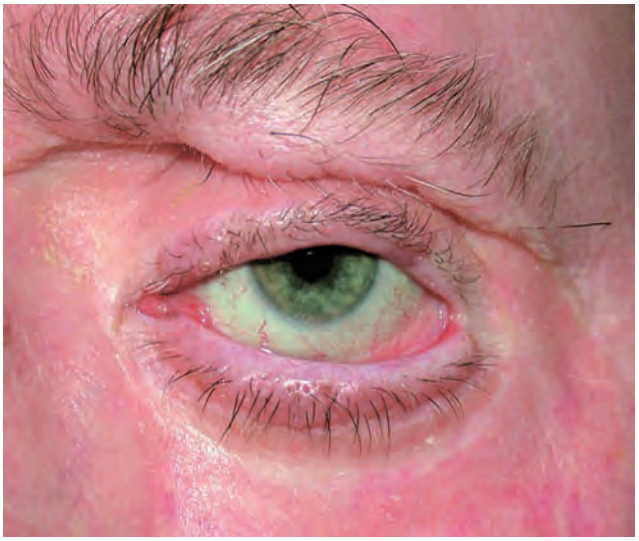


Fig. 1.64 Cicatricial ectropion

(Courtesy of A Pearson)

Fig. 1.65 Left facial palsy and severe paralytic ectropion

(Courtesy of A Pearson)

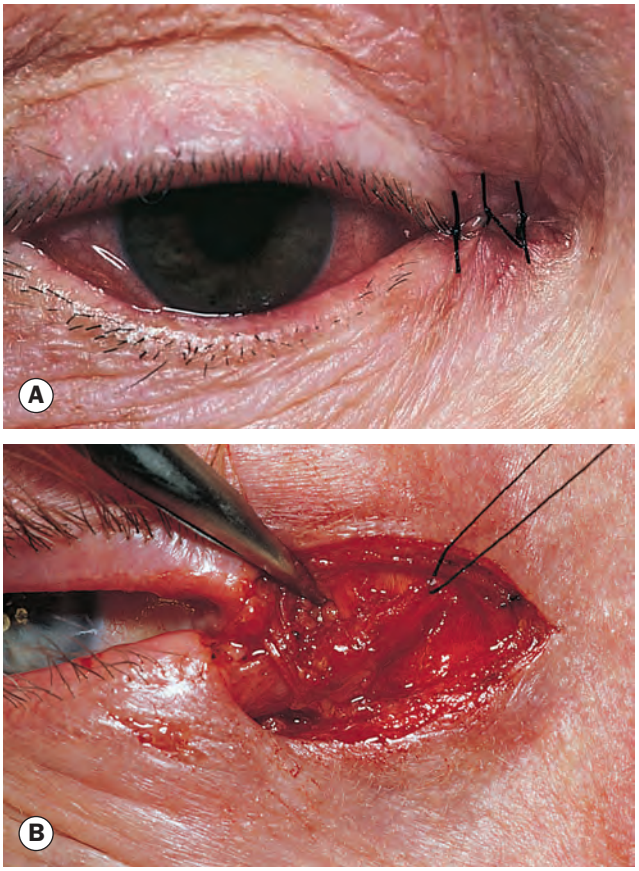


Fig. 1.66 Permanent treatment of paralytic ectropion. **(A)** Medial canthoplasty; **(B)** lateral canthal sling – refashioned canthal tendon from the lower lid is passed through a buttonhole in the tendon from the upper lid

(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001)*

Mechanical ectropion

Mechanical ectropion is caused by tumours on or near the lid margin that mechanically evert the lid. Treatment involves removal of the cause if possible, and correction of significant horizontal lid laxity.

ENTROPION

Involutional entropion

Introduction

Involutional (age-related) entropion affects mainly the lower lid. The constant rubbing of the lashes on the cornea in long-standing entropion (pseudotrichiasis – Fig. 1.67A) may cause irritation, corneal punctate epithelial erosions and, in severe cases, pannus formation and ulceration. Aetiological factors include:

- **Horizontal lid laxity** caused by stretching of the canthal tendons and tarsal plate.
- **Vertical lid instability** caused by attenuation, dehiscence or disinsertion of the lower lid retractors. Weakness of the latter is recognized by decreased excursion of the lower lid in downgaze.
- **Over-riding of the pretarsal** by the preseptal orbicularis during lid closure tends to move the lower border of the tarsal plate anteriorly, away from the globe, and the upper border towards the globe, thus tipping the lid inwards (Fig. 1.67B).
- **Orbital septum laxity** with prolapse of orbital fat into the lower lid.

Treatment

Temporary protection must be as short-term as possible; options include lubricants, taping, soft bandage contact lenses and orbicularis chemodenervation with botulinum toxin injection.

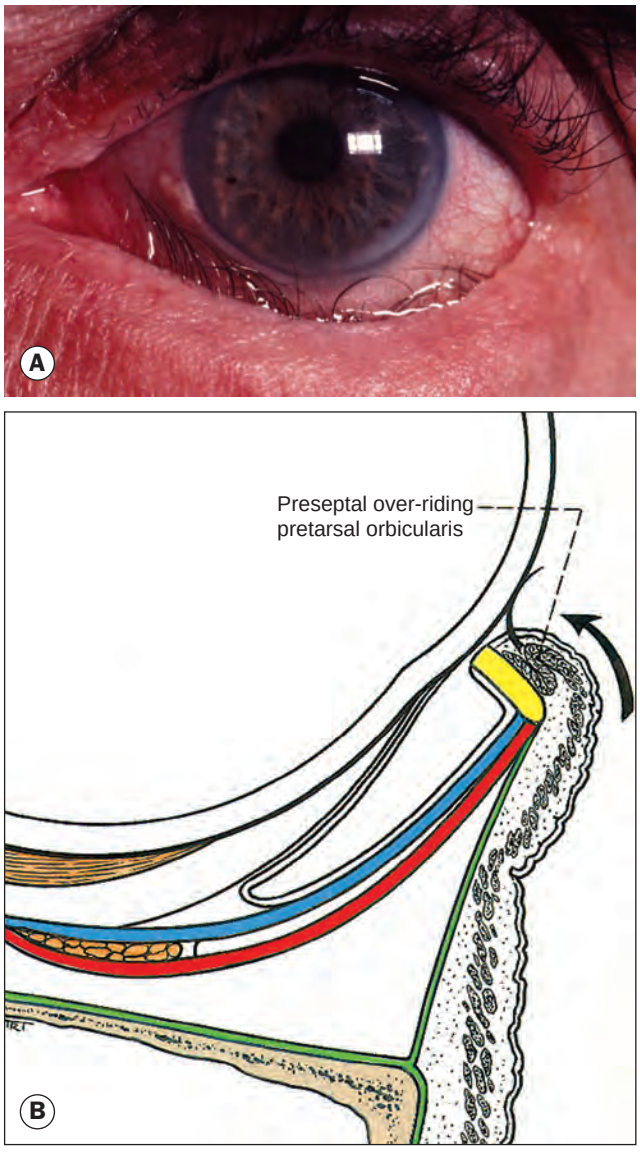


Fig. 1.67 **(A)** Involutional entropion and pseudotrichiasis; **(B)** preseptal orbicularis over-riding the pretarsal orbicularis

Surgical treatment aims to correct the underlying problems as follows:

- **Over-riding** and disinsertion
  - Transverse everting sutures prevent over-riding of the preseptal orbicularis. They are quick and easy to insert (Fig. 1.68), providing a correction typically lasting several months, and may be used in circumstances (e.g. a confused patient) where a more intricate procedure is not likely to be tolerated.
  - The Wies procedure gives a durable correction. It consists of full-thickness horizontal lid-splitting and insertion of everting sutures (Fig. 1.69). The scar creates a barrier between the preseptal and pretarsal orbicularis, and the everting suture fairly effectively transfers the pull of the lower lid retractors from the tarsal plate to the skin and orbicularis.
  - Lower lid retractor reinsertion (Fig. 1.70) involves direct exposure and advancement of the retractors as opposed to the less precise approach used in the Wies procedure. The subciliary skin incision used, and its repair, also create a barrier to over-riding of the preseptal orbicularis muscle. It can be performed as a primary treatment but may be reserved for recurrence.
- **Horizontal lid laxity** is usually present and can be corrected with a lateral canthal sling (tarsal strip – see Fig. 1.62) or, less commonly, a full-thickness lateral pentagon excision (see Fig. 1.63). Tightening serves also to retain the lid in apposition against the globe, preventing over-correction.

Cicatricial entropion

Scarring of the palpebral conjunctiva can rotate the upper or lower lid margin towards the globe. Causes include cicatrizing conjunctivitis, trachoma, trauma and chemical injuries. Temporary measures are similar to those listed for involutional entropion. Definitive surgical treatment of mild cases is by tarsal fracture (transverse tarsotomy) with anterior rotation of the lid margin, as for marginal entropion of the lower lid. Treatment of severe cases is difficult and is directed at replacing deficient or keratinized conjunctiva and replacing the scarred and contracted tarsal plate with composite grafts.

MISCELLANEOUS ACQUIRED DISORDERS

Varix

An eyelid varix (plural – varices) is a common lesion that may be mistaken for a naevus or haemangioma. A varix is commonly an

Fig. 1.68 Lid-everting sutures for entropion. **(A)** Three double-armed sutures are passed as shown; **(B)** sutures are tied; **(C)** schematic

(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001)*

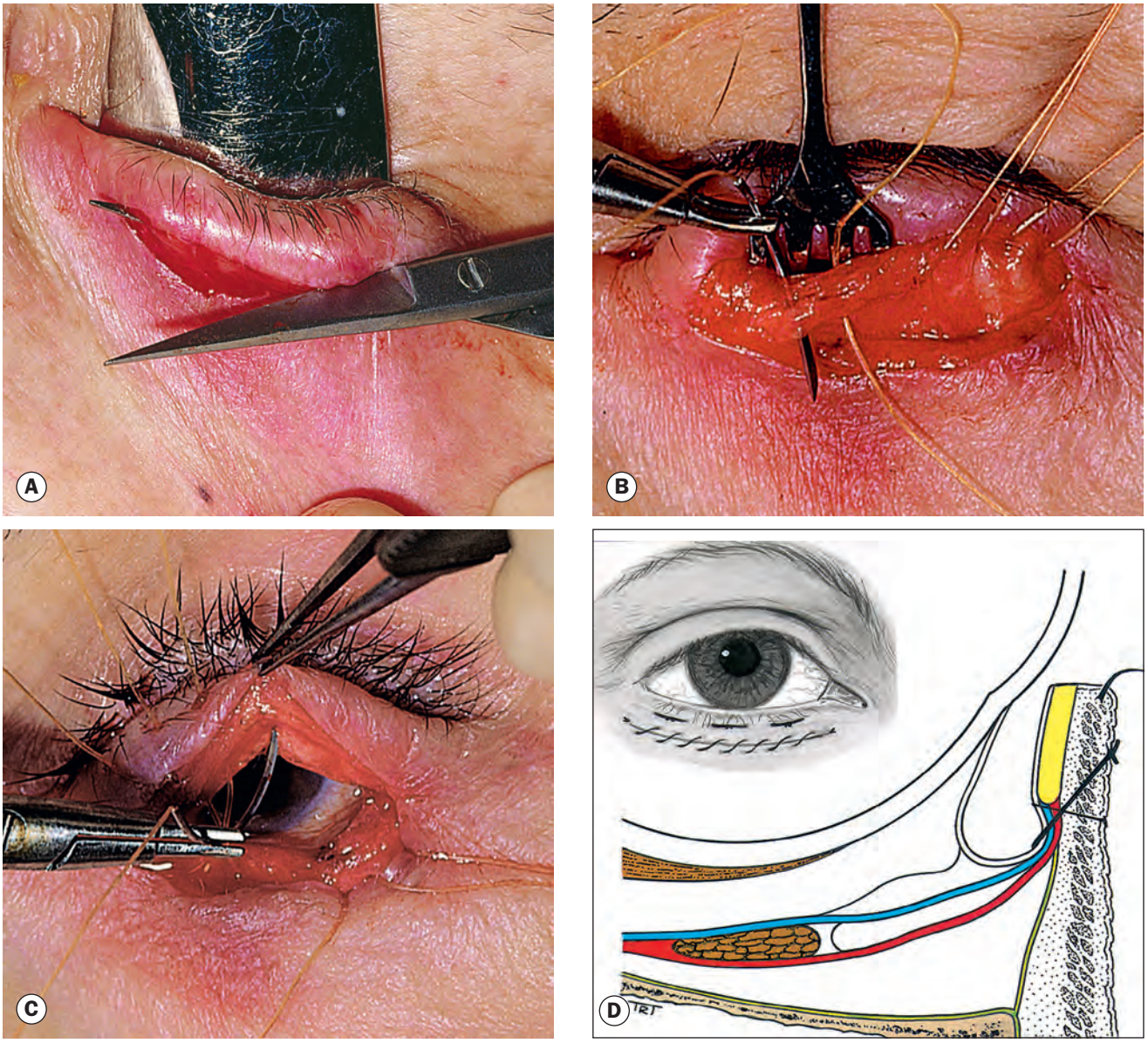


Fig. 1.69 Wies procedure for entropion. **(A)** Full-thickness incision; **(B)** sutures are passed through the conjunctiva and lower lid retractors; **(C)** sutures are passed anterior to the tarsal plate to exit inferior to the lashes; **(D)** schematic
(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001 – figs A–C)*

isolated lesion, but may be associated with orbital involvement (see Ch. 3). It appears as a dark red or purple subcutaneous compressible (unless thrombosed) lesion (Figs 1.71A and B), which in some cases becomes apparent only with a Valsalva manoeuvre (Figs 1.71C and D). It is clinically and histologically similar to a lymphangioma. Simple excision may be performed for diagnostic or cosmetic reasons; the possibility of orbital communication should be borne in mind during surgery.

Dermatochalasis

This is described in the discussion of pseudoptosis (above) and upper lid blepharoplasty (below).

Floppy eyelid syndrome

Introduction

Floppy eyelid syndrome (FES) is an uncommon unilateral or bilateral condition that is often overlooked as a cause of persistent ocular surface symptoms. It typically affects obese middle-aged and older men who sleep with one or both eyelids against the pillow, leading to pulling of the lid away from the globe; consequent nocturnal exposure and poor contact with the globe, often exacerbated by other ocular surface disease such as dry eye and blepharitis, result in chronic keratoconjunctivitis. Obstructive sleep apnoea (OSA) is strongly associated; OSA is linked to

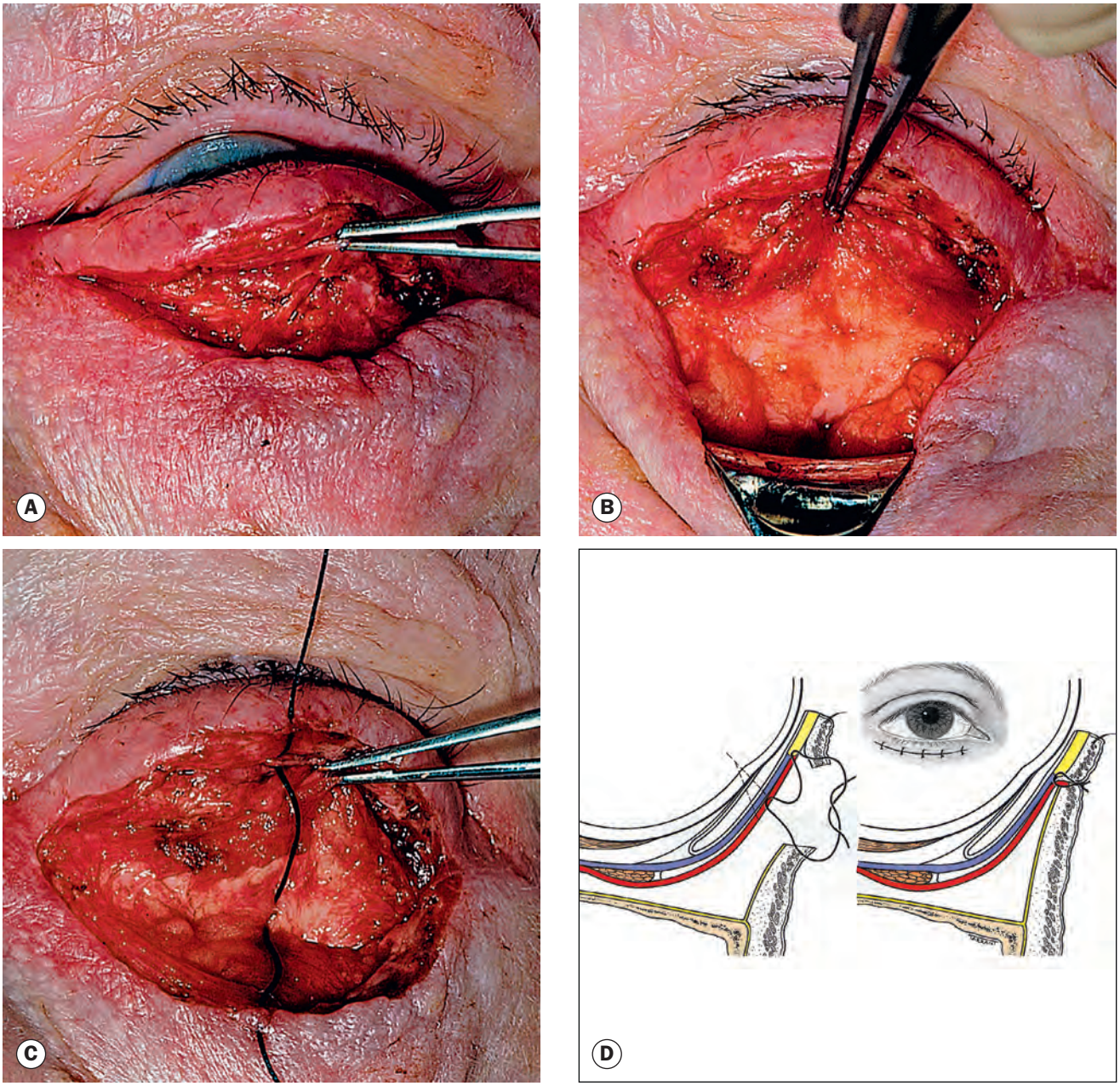


Fig. 1.70 Lower retractor reinsertion. **(A)** Incision to expose the lower border of the tarsal plate; **(B)** reflection of the orbital septum and fat pad to expose the lower lid retractors; **(C)** tightening of retractors by plication; **(D)** schematic
(Courtesy of AG Tyers and JRO Collin, from Colour Atlas of Ophthalmic Plastic Surgery, *Butterworth-Heinemann 2001)*

significant morbidity, including cardiopulmonary disease and subtle but irreversible mental dysfunction.

Diagnosis

- **The upper eyelid** is typically extremely lax, often with substantial excess loose upper lid skin (Fig. 1.72A). The tarsal plate has a rubbery consistency (Fig. 1.72B); the lid is very easy to evert (Fig. 1.72C), to fold and to pull away from the eye.
- **Papillary conjunctivitis** of the superior tarsal conjunctiva may be intense (Fig. 1.72D).
- **Keratopathy.** Punctate keratopathy, filamentary keratitis and superior superficial vascularization may be present.
- **Other findings** may include eyelash ptosis, lacrimal gland prolapse, ectropion and aponeurotic ptosis. Patients with both FES and OSA seem to have a considerably higher than average prevalence of glaucoma.
- **Investigation for OSA** should be considered in most cases of FES, particularly if the patient reports substantial snoring and/or excessive daytime sleepiness.

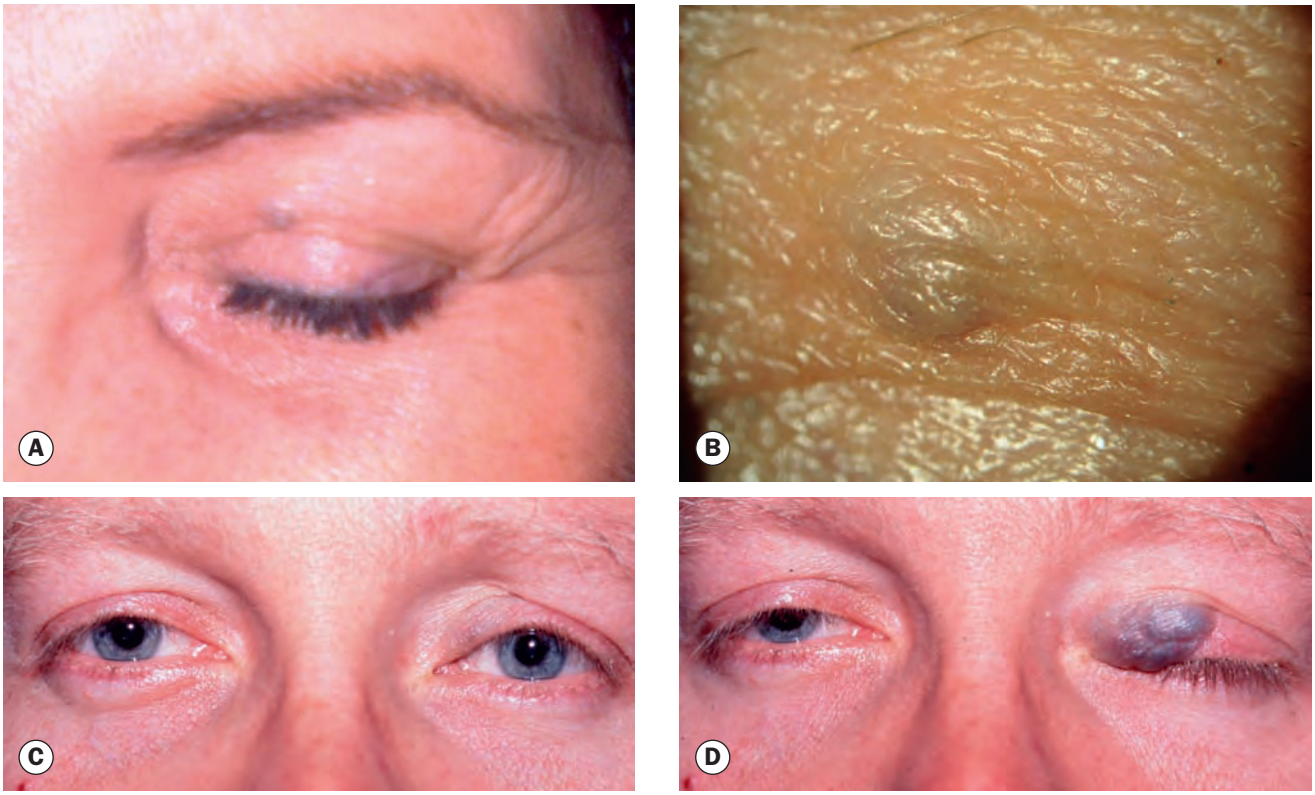


Fig. 1.71 Eyelid varices. **(A)** Typical appearance of a commonly seen small varix; **(B)** magnified view; **(C)** larger lesion, probably with orbital involvement, before Valsalva manoeuvre; **(D)** during Valsalva

(Courtesy of G Rose – figs C and D)

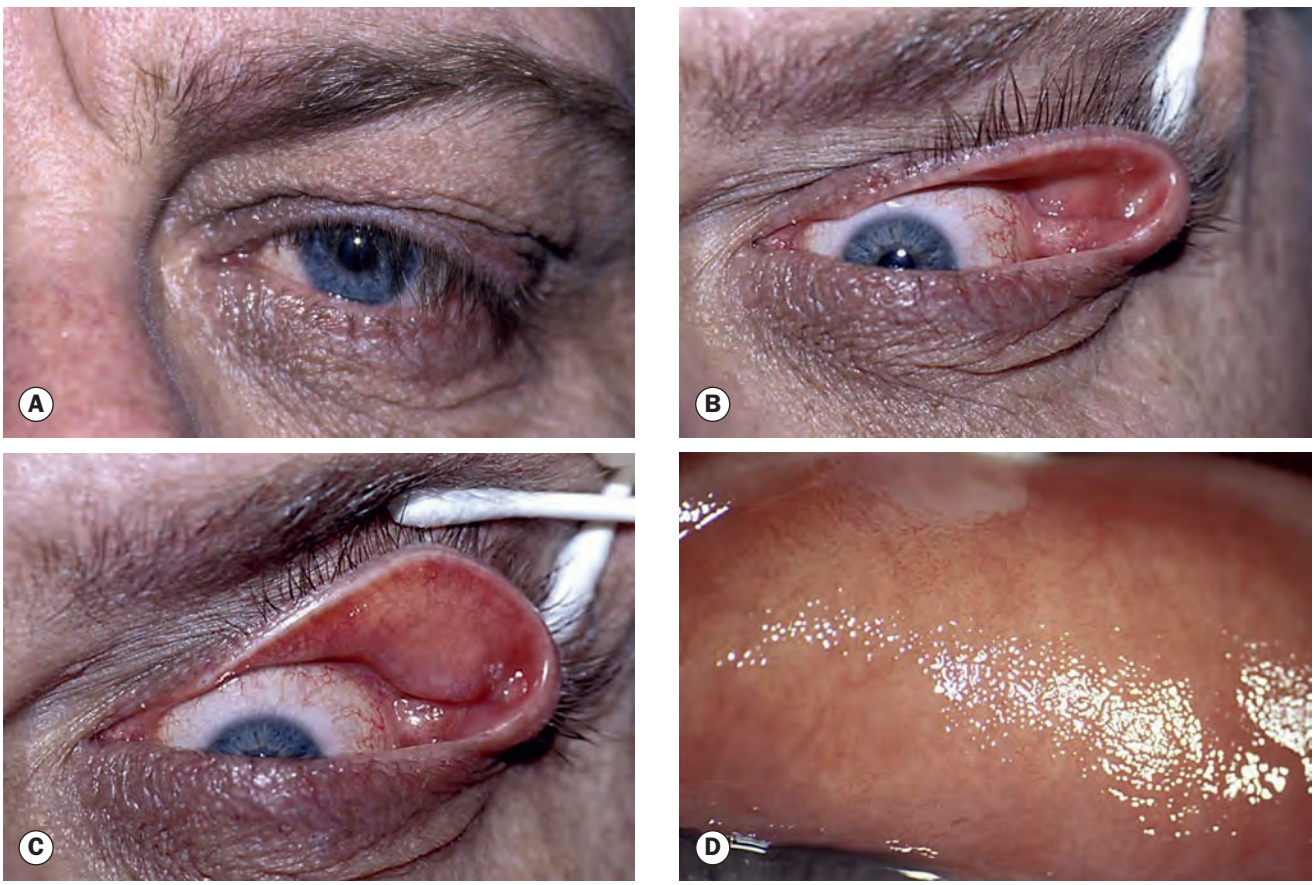


Fig. 1.72 Floppy eyelid syndrome. **(A)** Redundant upper lid skin; **(B)** loose and rubbery tarsal plates; **(C)** very easily everted eyelid; **(D)** superior tarsal papillary conjunctivitis

(Courtesy of C Barry)

Treatment

- Treatment of associated OSA is likely to be of benefit; overweight patients should be encouraged to lose weight.
- Mild cases may respond to lubrication together with nocturnal eye shield wear or taping of the lids.
- Moderate to severe cases require horizontal shortening to stabilize the lid and ocular surface and prevent nocturnal lagophthalmos; a pentagonal excision of 10 mm or more is taken from the junction of the lateral third and medial two-thirds of the upper lid.

Blepharochalasis

Blepharochalasis is an uncommon condition characterized by recurrent episodes of painless, non-pitting oedema of both upper lids which usually resolves spontaneously after a few days. Presentation is usually around puberty, episodes becoming less frequent with time. Eyelid skin becomes stretched and atrophic, characteristically said to resemble wrinkled cigarette paper; severe cases may give rise to stretching of the canthal tendons and levator aponeurosis resulting in ptosis (Fig. 1.73), and lacrimal gland prolapse may occur. A hypertrophic form with orbital fat herniation and an atrophic form with absorption of orbital fat have been described. The differential diagnosis includes similarly episodic conditions, particularly drug-induced urticaria and angioedema. Treatment involves blepharoplasty for redundant upper lid skin, and correction of ptosis.

Eyelid imbrication syndrome

Eyelid imbrication syndrome is an uncommon and frequently unrecognized disorder in which the upper lid overlaps the lower on closure so that the lower lashes irritate the superior marginal tarsal conjunctiva.

It may be unilateral or bilateral and the major symptom is ocular irritation. It can be acquired, commonly associated with floppy eyelid syndrome, or – very rarely – congenital; occasionally it may follow lower lid tarsal strip surgery. Associated signs include superior tarsal papillary conjunctivitis and rose Bengal staining of the superior marginal conjunctiva. Definitive treatment consists of upper lid pentagon resection and/or lateral canthal tightening.

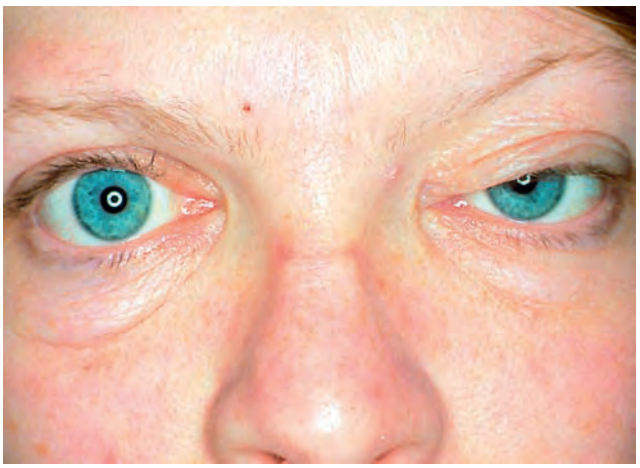


Fig. 1.73 Blepharochalasis – left aponeurotic ptosis and thinned upper lid skin

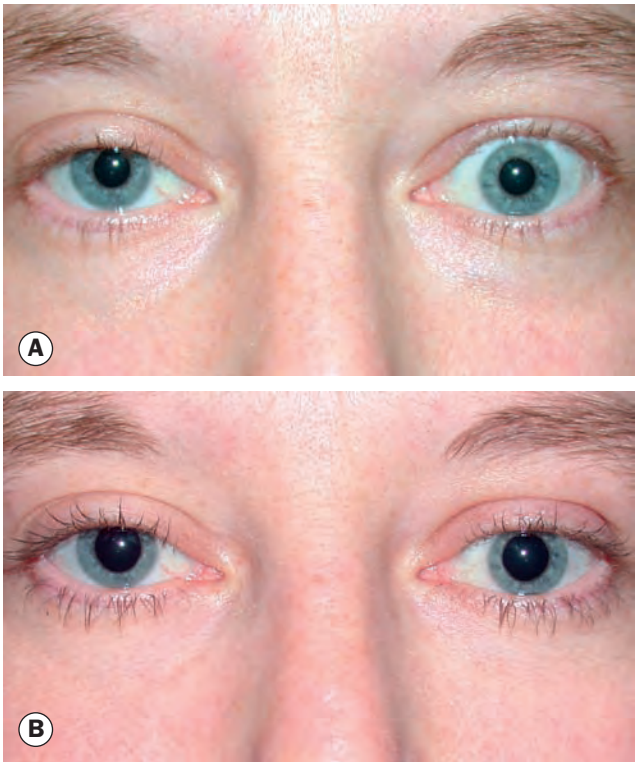


Fig. 1.74 **(A)** Left lid retraction in thyroid eye disease; **(B)** following Müller muscle recession
(Courtesy of A Pearson)

Upper lid retraction

Upper lid retraction is suspected when the upper lid margin is either level with or above the superior limbus (Fig. 1.74A); the causes are listed in Table 1.5. Where there is no loss or tightness of the upper eyelid skin, retraction is corrected by surgical release of the eyelid retractors, usually via a transconjunctival posterior approach. Mild retraction may be treated with Müller muscle recession (Fig. 1.74B). Moderate to severe retraction may require levator aponeurosis recession.

Lower lid retraction

Inferior scleral show may be physiological in patients with large eyes or shallow orbits, but is commonly involutional or secondary to some of the conditions in Table 1.5. It may follow lower lid blepharoplasty, when aggressive upward massage of the lid for 2 or 3 months may be curative for minor degrees. In other cases, a tarsal strip operation may raise the lid slightly, but when moderate elevation is required inferior retractor recession with a posterior lamellar spacer is likely to be necessary; more aggressive procedures have been described for severe cases.

Table 1.5 Causes of lid retraction

1. Thyroid eye disease
2. Neurogenic • Contralateral unilateral ptosis (Fig. 1.75A) • Unopposed levator action due to facial palsy • Third nerve misdirection • Marcus Gunn jaw-winking syndrome • Collier sign of the dorsal midbrain (Parinaud syndrome – Fig. 1.75B) • Infantile hydrocephalus (setting sun sign – Fig. 1.75C) • Parkinsonism (Fig. 1.75D) • Sympathomimetic drops
3. Mechanical • Surgical over-correction of ptosis • Scarring of upper lid skin
4. Congenital • Isolated • Duane retraction syndrome • Down syndrome • Transient 'eye popping' reflex in normal infants
5. Miscellaneous • Prominent globe (pseudo-lid retraction) • Uraemia (Summerskill sign) • Idiopathic

COSMETIC EYELID AND PERIOCULAR SURGERY

Involutional changes

Involutional (age-related) changes around the eyes can lead to functional and cosmetic concerns that may require treatment.

- Reduction in cutaneous elasticity and thickness results in loose, wrinkled skin.
- Weakening of the orbital septum may lead to orbital fat prolapse.
- Thinning and stretching of the canthal tendons, levator aponeurosis and lower lid retractors may cause eyelid laxity and ptosis.
- Atrophy of orbital and eyebrow fat pads can give enophthalmos and eyebrow sagging.
- Weakening of the frontalis muscle and epicranial aponeurosis may cause descent of the eyebrows and increasing looseness of upper eyelid skin.
- Thinning and stretching of midfacial support leads to descent with formation of a tear trough depression and exacerbation of lower eyelid changes.
- Thinning and resorption of periorbital bone exacerbates the appearance of surplus overlying tissues.

Non-surgical techniques

Botulinum toxin injection to periocular muscles

Botulinum toxin injection can be used to reduce wrinkling, particularly for 'crows' feet' at the lateral canthus and for glabellar frown lines, and 'brow lift' by a reduction in the action of brow depressors. Complications include temporary ptosis, lagophthalmos, ectropion and diplopia.

Tissue fillers

These are used to address age-related wrinkles, and less commonly defects from other causes such as trauma. Complications include hypersensitivity reactions.

- **Hyaluronic acid** is the most commonly used tissue filler, and can be used to temporarily fill in hollows and replace lost volume. They are injected deep to orbicularis and the effects generally last 3–12 months depending on the agent used.
- **Autologous fat** gives a more permanent replacement.
- **Others** include collagen, microspheres of calcium hydroxyapatite and synthetic fillers.

Skin resurfacing

Removal of the superficial layers of the skin, by chemical peels or laser, can lead to a reduction in wrinkling, increased evenness of pigmentation, removal of blemishes and improved texture by generating new epidermis and increasing collagen production in the dermis.

Surgical techniques

Upper eyelid blepharoplasty

Upper eyelid involutional changes are characterized by surplus upper eyelid skin (dermatochalasis) that leads to baggy lids with indistinct creases and pseudo- or mechanical ptosis. It may cause a heavy sensation around the eyes, brow ache and, in more advanced cases, obstruction of the superior visual field (Fig. 1.76A). Upper lid blepharoplasty (Fig. 1.76B) is effective for the removal of surplus skin and can be combined with reduction of the superior orbital fat pads. Care must be taken prior to surgery to look for ptosis of the eyelid or eyebrow and ocular surface dryness. Complications include removal of excess skin leading to lagophthalmos and corneal drying, and removal of excess orbital fat leading to an unattractive hollowed out upper eyelid sulcus.

Lower eyelid blepharoplasty

Lower lid involutional changes are characterized by excess skin and/or prolapsed orbital fat (Fig. 1.77A); blepharoplasty can address these (Fig. 1.77B).

- **Anterior approach.** Where there is excess skin an anterior approach is used to raise a skin/muscle flap that can be lifted and re-draped on the lid with the surplus removed. At the same time the inferior orbital fat pads can be reduced by a small incision through the septum.
- **Posterior approach.** Bulging of the lower eyelid fat pads without eyelid laxity or surplus skin is best reduced by a

Fig. 1.75 Causes of lid retraction. **(A)** Unilateral myasthenic ptosis with contralateral lid retraction; **(B)** Collier sign; **(C)** 'setting sun' sign in infantile hydrocephalus; **(D)** parkinsonism
(Courtesy of R Bates – fig. C)

posterior, transconjunctival approach. Complications include lower eyelid retraction, contour abnormalities (particularly lateral drooping), and frank ectropion.

Brow ptosis correction

Brow ptosis frequently accompanies dermatochalasis (Fig. 1.78A) and may also follow facial nerve palsy or localized trauma. Lifting of the brow needs to precede or occasionally be combined with upper lid blepharoplasty.

- **Direct brow lift.** An incision is made above the eyebrow hairs and an ellipse of skin removed (Fig. 1.78B).
- **Endoscopic brow lift.** Small incisions within the hair-line enable endoscopic elevation of the whole forehead tissues and release at the eyebrow periosteum to allow lifting of the eyebrows through sutures supported on frontal bone anchors within the hair-line.

CONGENITAL MALFORMATIONS

Epicanthic folds

Epicanthic folds are bilateral vertical folds of skin that extend from the upper or lower lids towards the medial canthi. They may give rise to a pseudoexotropia. The folds may involve the upper or lower lids or both; lower lid folds extending upwards to the medial canthal area (epicanthus inversus – Fig. 1.79A) are associated with the blepharophimosis syndrome. Treatment is by V–Y (Fig. 1.79B) or Z-plasty.

Telecanthus

Telecanthus is an uncommon condition that may occur in isolation or in association with blepharophimosis and some systemic

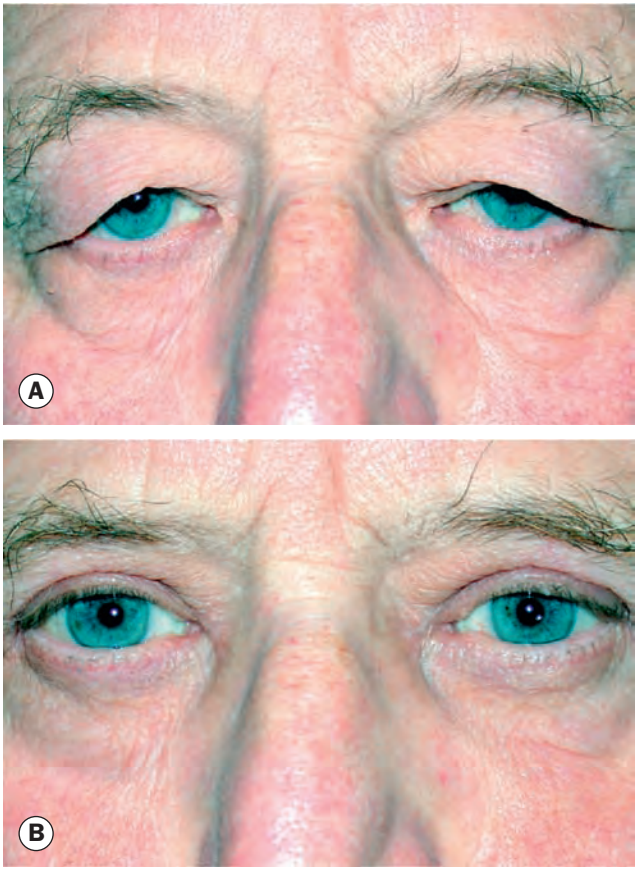


Fig. 1.76 **(A)** Severe dermatochalasis causing reduction of upper visual field; **(B)** appearance following surgery
(Courtesy of A Pearson)

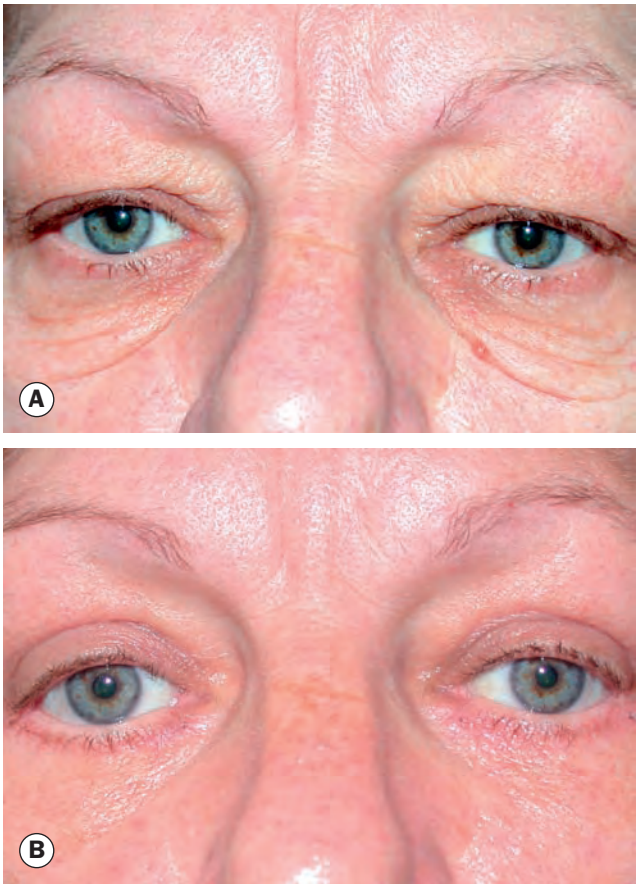


Fig. 1.77 **(A)** Mild dermatochalasis and excess lower lid skin; **(B)** appearance following upper and lower lid blepharoplasty
(Courtesy of A Pearson)

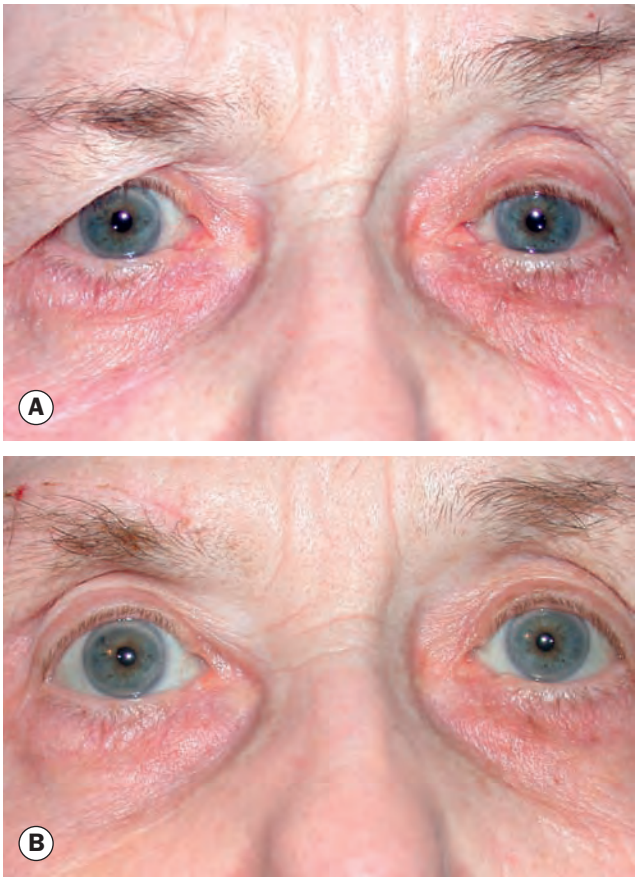


Fig. 1.78 **(A)** Right brow ptosis and dermatochalasis; **(B)** following direct brow-lift
(Courtesy of A Pearson)

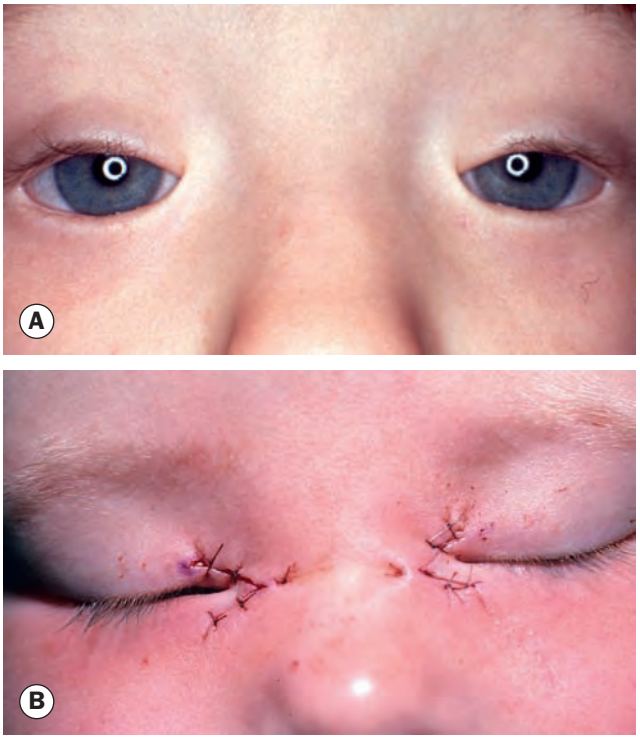


Fig. 1.79 Epicanthus inversus. **(A)** Preoperative appearance; **(B)** immediately after V–Y plasty
(Courtesy of R Bates – fig. B)

Fig. 1.80 Telecanthus

syndromes. It consists of increased distance between the medial canthi due to abnormally long medial canthal tendons (Fig. 1.80). It should not be confused with hypertelorism in which there is wide bony separation of the orbits. Treatment involves shortening and refixation of the medial canthal tendons to the anterior lacrimal crest, or insertion of a trans-nasal suture.

Blepharophimosis, ptosis and epicanthus inversus syndrome

Blepharophimosis, ptosis and epicanthus inversus syndrome (BPES) is a complex of eyelid malformations consisting of moderate to severe symmetrical ptosis with poor levator function, telecanthus, epicanthus inversus (see Fig. 1.79A), manifesting with small palpebral fissures (Fig. 1.81). Other minor facial anomalies

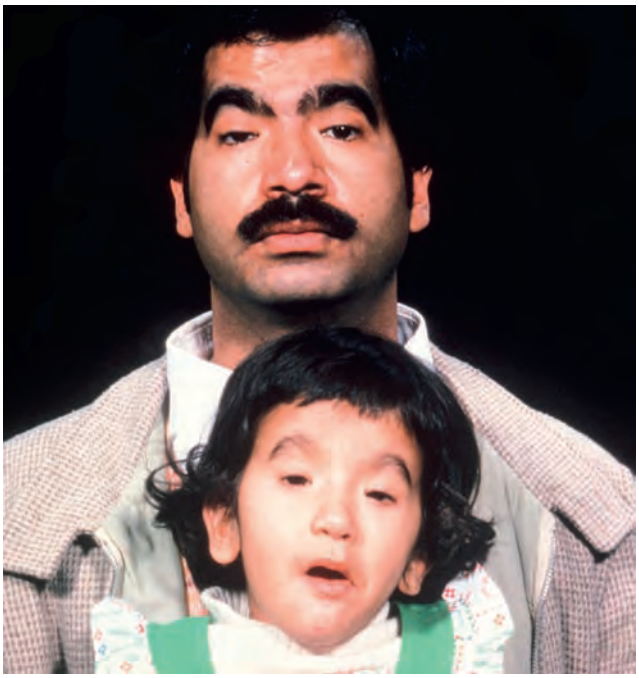


Fig. 1.81 Blepharophimosis ptosis and epicanthus inversus syndrome

are commonly present. Inheritance is usually autosomal dominant; both BPES type I (with premature ovarian failure) and BPES type II (without premature ovarian failure) are caused by mutations in the *FOXL2* gene on chromosome 3. Treatment initially involves correction of epicanthus and telecanthus, followed later by bilateral frontalis suspension. It is also important to treat amblyopia, which is present in about 50%.

Epiblepharon

Epiblepharon comprises an extra horizontal fold of skin stretching across the anterior lid margin; it is very common in individuals of Eastern Asian ethnicity. The lashes are directed vertically, especially in the medial part of the lid (Figs 1.82A and B). When the fold of skin is pulled down the lashes turn out and the normal location of the lid becomes apparent (Fig. 1.82C); it should not be confused with the much less common congenital entropion (see next). Treatment is not required in the majority of Caucasians because spontaneous resolution with age is usual. Persistent cases may be treated surgically.

Congenital entropion

Upper lid entropion is usually secondary to the mechanical effects of microphthalmos, which cause variable degrees of upper lid inversion. Lower lid entropion (Fig. 1.83) is generally caused by maldevelopment of the inferior retractor aponeurosis. Treatment involves the excision of a strip of skin and muscle, and fixation of the skin crease to the tarsal plate (Hotz procedure).

Coloboma

A congenital coloboma is an uncommon, unilateral or bilateral, partial- or full-thickness eyelid defect. It occurs when eyelid development is incomplete, due to either failure of migration of lid ectoderm to fuse the lid folds or to mechanical forces such as amniotic bands. Colobomata elsewhere in the eye, as well as a range of other associations, may be present. The treatment of small defects involves primary closure, while large defects require skin grafts and rotation flaps.

- **Upper lid colobomas** occur at the junction of the middle and inner thirds (Fig. 1.84A); relatively strong associations include cryptophthalmos (see below), facial abnormalities and Goldenhar syndrome.
- **Lower lid colobomas** occur at the junction of the middle and outer thirds (Fig. 1.84B) and are frequently associated with systemic conditions.
- **Treacher Collins syndrome** (mandibulofacial dysostosis) is a genetically heterogeneous condition characterized by malformation of derivatives of the first and second branchial arches, principally mandibular and ear anomalies. Lower eyelid coloboma is a feature; ocular anomalies also described include slanted palpebral apertures, cataract, microphthalmos and lacrimal atresia.

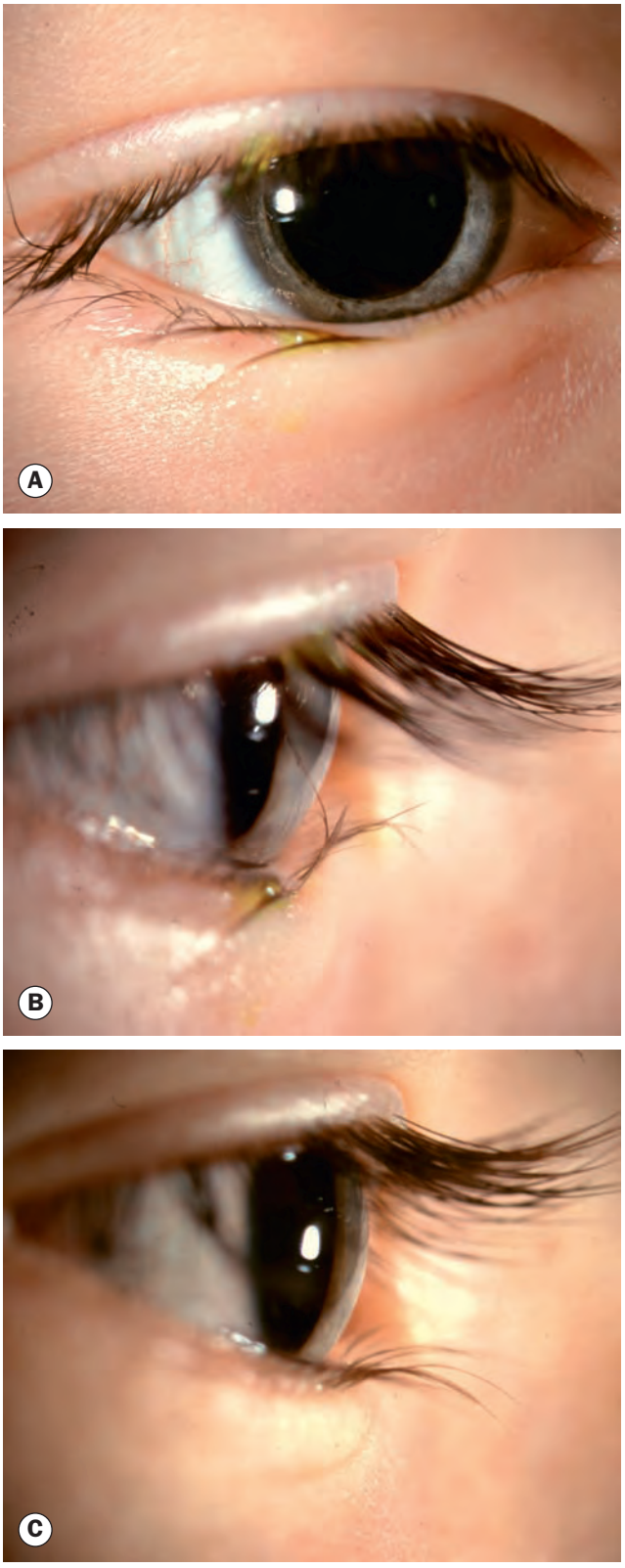


Fig. 1.82 **(A)** Epiblepharon; **(B)** lashes pointing upwards; **(C)** normal position of lashes following manual correction

Fig. 1.83 Congenital lower lid entropion

Cryptophthalmos

Cryptophthalmos is a rare congenital anomaly in which the eyelids are absent, replaced by a continuous layer of skin.

- **Complete cryptophthalmos.** A microphthalmic eye (Fig. 1.85A) is covered by a fused layer of skin with no separation between the lids.
- **Incomplete cryptophthalmos** is characterized by rudimentary lids and microphthalmos (Fig. 1.85B).
- **Fraser syndrome** is a dominantly inherited condition in which cryptophthalmos is a common finding; other features can include syndactyly, urogenital and craniofacial anomalies.

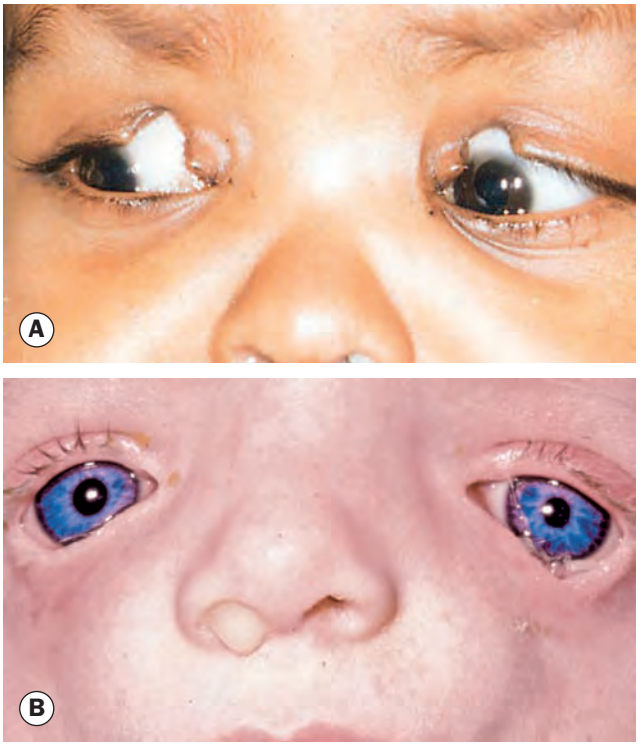


Fig. 1.84 **(A)** Upper lid colobomas; **(B)** lower lid colobomas in Treacher Collins syndrome
(Courtesy of U Raina – fig. A)

Fig. 1.85 Cryptophthalmos. **(A)** Complete; **(B)** incomplete
(Courtesy of D Meyer – fig. A)

Euryblepharon

Euryblepharon refers to horizontal enlargement of the palpebral fissure with associated lateral canthal malposition and lateral ectropion (Fig. 1.86); lagophthalmos and exposure keratopathy may result.

Microblepharon

Microblepharon is characterized by small eyelids, often associated with anophthalmos (Fig. 1.87).

Fig. 1.86 Euryblepharon
(Courtesy of D Taylor and C Hoyt, from Pediatric Ophthalmology and Strabismus, *Elsevier 2005)*

Fig. 1.87 Microblepharon associated with anophthalmos

Ablepharon

Ablepharon consists of deficiency of the anterior lamellae of the eyelids (Fig. 1.88A); treatment involves reconstructive skin grafting. Ablepharon-macrostomia syndrome is characterized by an enlarged fish-like mouth (Fig. 1.88B), ear, skin and genital anomalies.

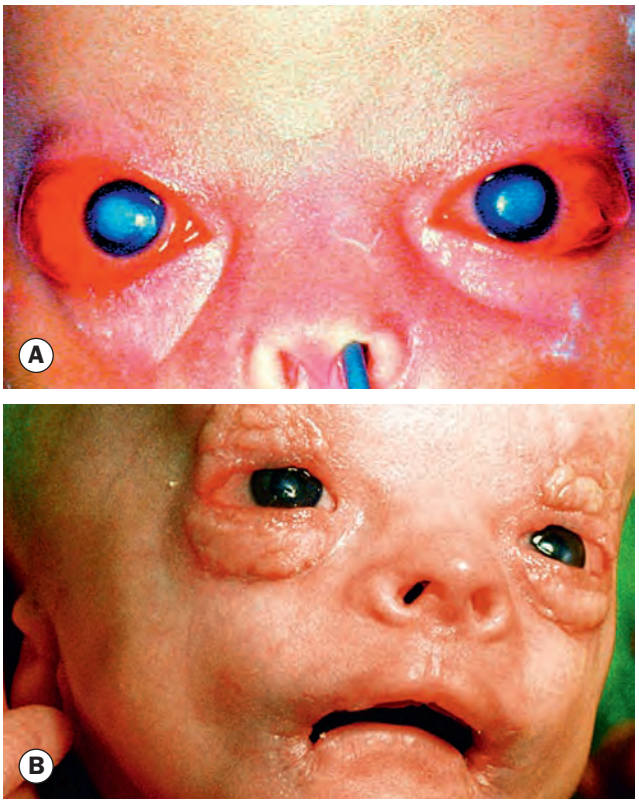


Fig. 1.88 **(A)** Ablepharon; **(B)** following reconstruction – note enlarged fish-like mouth
(Courtesy of D Taylor and C Hoyt, from Pediatric Ophthalmology and Strabismus, *Elsevier 2005 – fig. A; H Mroczkowska – fig. B)*

Congenital upper lid eversion

Congenital upper lid eversion is a rare condition more frequently seen in infants of Afro-Caribbean origin, in Down syndrome and in congenital ichthyosis (collodion skin disease – Fig. 1.89). It is typically bilateral and symmetrical. It may resolve spontaneously with conservative treatment or require surgery.

Fig. 1.89 Congenital upper lid eversion in a patient with ichthyosis
(Courtesy of D Meyer)

Fig. 1.90 Ankyloblepharon filiforme adnatum
(Courtesy of D Taylor and C Hoyt, from Pediatric Ophthalmology and Strabismus, *Elsevier 2005)*

Ankyloblepharon filiforme adnatum

In ankyloblepharon filiforme adnatum the upper and lower eyelids are joined by thin tags (Fig. 1.90); most cases are sporadic. Treatment involves transection with scissors; anaesthesia is not required.